NOMENCLATURE

*and*

CRITERIA FOR DIAGNOSIS

*of*

# DISEASES OF THE HEART AND BLOOD VESSELS

# NOMENCLATURE AND CRITERIA

FOR DIAGNOSIS OF

# DISEASES OF THE HEART AND BLOOD VESSELS

BY


THE CRITERIA COMMITTEE

OF THE

NEW YORK HEART ASSOCIATION INC

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*The Committee requests that you make the following corrections in this printing*

PAGE 139 line 16 should read
the designation $V_{4R}$

PAGE 172 first footnote line 5 should read
in microvolt seconds ($\mu$vs)

PAGE 175 Table I under P wave mean value
of $aV_R$ should be −1 09

PAGE 216 Fig 12 end of line 1 should read
a thick layer of fibrin beneath

PAGE 303 insert at bottom of page
g Other toxic agent 190–3

PAGE 336 add
g Other toxins

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THIS BOOK IS DEDICATED

TO THE MEMORY OF

JOHN WYCKOFF, M.D.

who did much to build up
the New York Heart Association

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# PREFACE

THE FIFTH EDITION of the criteria has been expanded to include a section on diseases of the peripheral vessels. It also has afforded an opportunity to introduce such changes of opinion and viewpoint as have occured in the last ten years. The more inclusive definition of disease introduced in the fourth edition has been retained so that not only structural or organic changes but also functional or physiological disturbances are included as forms of diseases. This has particular importance in considering the Functional Capacity and the Therapeutic Classification of cardiac patients as well as in the diseases of the blood vessels.

Certain features seem to warrant special mention. The criteria for the diagnosis of rheumatic fever, an important etiologic factor in cardiac disease, have been amplified and made more exacting. The suggestions of the previous editions as to a standard terminology for the description of heart sounds and murmurs have been revised and we hope improved. Considerable confusion continues to arise from the lack of standard terms for these diagnostic sounds. The term "innocent murmur" has been introduced to designate those murmurs not believed to be the result of cardiac disease. Because of the prognostic implication this seems an improvement on previous terms such as functional, accidental or unknown. The criteria for the diagnosis of congenital anomalies have been greatly expanded with the invaluable help of Dr Gertrude H. B. Nicolson. Surgery offers so much in many of these cases that we must make every effort to obtain an exact and complete diagnosis.

A new discussion of cardiac insufficiency presents the most recent thought on this important physiological disturbance. The retention of this term has seemed preferable to the use of "cardiac failure" because the latter term so often is applied specifically to the terminal stages of the process. It seemed better to continue to use the newer term rather than to attempt to re-educate the physician in a broader and less specific use of the older one.

The Therapeutic Classification which first appeared in the previous edition has proved its usefulness in the clinic as an indication of the amount of physical activity that may be allowed to the patient. It is a useful addition to the rating of Functional Capacity, which is determined only by the limitation of activity enforced by the cardiac disease. Since this limitation does not always parallel the degree to which

the patient's activity should be restricted, it cannot afford the sole guide to such management.

In response to a request by the Committee on Rheumatic Fever and Congenital Heart Disease of the American Heart Association it has been decided to discontinue the use of the terms Possible Heart Disease and Potential Heart Disease. The new terminology, though it may seem strange to those accustomed to the old one, has the advantage of being more specific and of avoiding the use of the term Heart Disease in connection with patients not known to be suffering from this condition.

The Roentgenological Section has been extensively re-written and a number of excellent new illustrations have been furnished by Dr. Schwedel. The Electrocardiographic Section has important new material on criteria for adequate recording devices, criteria for techniques, criteria for nomenclature of the waves and many new illustrations. In the Criteria for Interpretation all leads generally available—bipolar and unipolar extremity, precordial and esophageal—are included in defining the electrocardiographic picture. These criteria should enable a physician to designate exactly any electrocardiographic phenomenon. They are not to be considered as a set of rules for the differentiation of the normal from the abnormal electrocardiogram. The Committee has not presented in this section a description of specific diagnostic patterns but whenever the electrocardiographic features are sufficiently characteristic to be used as one of the criteria for clinical diagnosis these patterns are described in the appropriate part of the text of the Anatomical Section.

The Criteria for the Pathological Diagnosis of Diseases of the Heart and Great Vessels was published as an appendix in the previous edition, having been prepared under the auspices of the Research Committee. In this edition it has been made an integral part of the text and the committee responsible has become a subcommittee of the Criteria Committee. This has made possible a better coordination of the diagnoses of the clinical section on Anatomical Diagnosis with analogous parts of the Pathological Diagnosis. The chapter on congenital anomalies has been rewritten by Dr. Henry W. Edmonds whose approach to the subject differs somewhat from that presented by Dr. Nicolson in the Anatomical Section. The former follows anatomical subdivisions and the latter clinical. Thanks are due to Dr. Maurice Lev who has helped in the revision of the chapter on the conduction system. New data have been added on Injuries of the Heart and Great Vessels and there are notes on Aims and Procedures in the autopsy study of congenital anomalies of the heart. The illustrations have been improved

by replacements and additions We wish to thank The C V Mosby Co Grune and Stratton and the American Medical Association for permission to reproduce Figures 21 22 and 23 37 and 38 and 39 respectively We are also indebted to the Armed Forces Institute of Pathology for the privilege of reproducing Figures 26 30 31 32 and 33 This section has aroused great interest among pathologists and it is hoped that the terminology and criteria suggested will lead to more uniformity of thought and less possibility of misunderstanding in this field as has been the case with those using the clinical criteria

A wholly new section has been added on the subject of peripheral vascular diseases The subcommittee under the direction of Dr Irving S Wright has done pioneer work in planning a nomenclature for these diseases They have not found it satisfactory to treat them as has been found so useful in dealing with diseases of the heart considering the diseases as a group of physiological and structural phenomena resulting from the effect of various etiological agents The nomenclature presented is more an analysis of the subject than a list of diagnostic titles This was considered by the subcommittee as the best method of presenting to the profession this group of diseases which has only recently come to be viewed as an entity It is hoped that with further consideration of the subject these diseases also may be found to fit into the generally used scheme of the nomenclature of disease

The appropriate code numbers of the Standard Nomenclature for Diseases and Operations sponsored by the American Medical Association have been indicated when possible in both the Cardiac and the Peripheral Vascular sections We are indebted to Adeline C Hayden Associate Editor of the recent edition of this work for help in selecting the proper code numbers

Thanks are due to all the members of the subcommittees who have helped in the writing of each of the sections The viewpoints brought to this work by each one of them has been a stimulus to the central committee and an important factor in maintaining the high standard of the text The text prepared by the subcommittees on Etiological Anatomical and Physiological Diagnosis and on Functional Capacity and Therapeutic Classification was revised in detail and edited by the Criteria Committee The text of the subcommittees on Electrocardiographic and Roentgenologic Diagnosis and on Peripheral Vascular Diseases was revised and edited by the Chairman of the criteria committee in consultation with the chairman of the appropriate subcommittee

HAROLD E B PARDEE M D
Chairman

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## SECTION ON PATHOLOGICAL DIAGNOSIS

# NOMENCLATURE FOR CARDIAC DIAGNOSIS

## ON THE USE OF CODE NUMBERS

Where dots appear in the etiological or anatomical series of numbers of the Standard Nomenclature they are to be replaced by the appropriate numeral to specify the exact detail of diagnosis appropriate to the particular case.

In the Anatomical Section several etiological numbers are sometimes shown. These are merely examples and not intended to exclude other etiological diagnoses.

The physiological diagnosis is considered by the Standard Nomenclature to be a manifestation. The numerals then form a third group following those indicating anatomical and etiological diagnosis. If the cause is unknown, the etiological diagnosis may be expressed by x followed by the last two numbers of the physiological diagnosis, e.g. x25 to indicate transient atrial fibrillation of unknown cause.

# NOMENCLATURE FOR CARDIAC DIAGNOSIS

DISEASES of the heart are here considered to include not only the structural changes found in the heart, pericardium and adjacent arterial structures but also such disturbances of the functions of the heart as are manifested by arrhythmia by its ability to perform its function as a pump and by its tendency to give rise to abnormal sensations This broader definition of disease brings within the scope of this nomenclature a number of disturbances that formerly had to be dismissed as functional even through their consequences might have been definitely disabling Obviously the usefulness of the nomenclature is increased by this change

A complete diagnosis should include one or more titles from each of the main headings of this nomenclature There should be a statement as to the etiology of the disease If structural changes are discovered they should be named or there should be a statement that there is no structural disease There should be a statement indicating the cardiac mechanism and any disturbance of cardiac physiology which may have arisen particularly a diagnosis to indicate the patient s symptomatology A diagnosis of the cardiac functional capacity and a statement of the patient s therapeutic classification completes the list Thus a comprehensive diagnosis will demand a careful consideration of every aspect of the case and will afford a sound basis for prognosis and treatment

Certain patients may have symptoms or abnormal physical signs and yet it may not be possible to make a diagnosis of structural disease or of a disturbance of cardiac physiology These should be retained for further consideration with the title Undiagnosed Manifestation Patients without structural or physological disturbances of the heart but who have had some other disease capable of causing heart disease may be retained for further observation under the diagnosis No Heart Disease Predisposing Etiological Factor and there should be a statement of the etiological factor

# ETIOLOGICAL DIAGNOSIS

| | | Code Numbers* |
|---|---|---|
| 1 | Anemia | –540 |
| 2 | Animal parasites | –2 |
| 3 | Arteriosclerosis | –912 |
| 4 | Bacterial infection** | –100 |
| 5 | Congenital anomaly | –0 |
| 6 | Hypertension | –533 |
| 7 | Hyperthyroidism | –771 |
| 8 | Hypothyroidism | –772 |
| 9 | *Neoplasm* | –8 |
| 10 | Neurocirculatory asthenia (effort syndrome) | –580 |
| 11 | Other etiological factors *(to be specified)* | |
| 12 | Other infections** | –1 |
| 13 | Pulmonary arterial hypertension | –535 |
| 14 | Reflex disturbance of the heart | –582 –584 –589 –590 |
| 15 | Rheumatic fever** | –932 |
| 16 | Syphilis** | –117 |
| 17 | Thiamin deficiency | –7621 |
| 18 | Toxic agent | –3 |
| 19 | Trauma | –4 |
| 20 | Unknown | –x00–functional reaction alone manifest<br>–900–structural reaction alone manifest |

*Code Numbers of Standard Nomenclature of Diseases and Operations. Richard J. Plunkett, M.D., and Adeline C. Hayden for the American Medical Association. Blakiston, 1952.

**When one of these diagnoses is used it should be stated, if possible, whether the etiological factor is active or is inactive—e.g., Rheumatic fever, inactive; Bacterial infection, streptococcus viridans, active.

# ANATOMICAL DIAGNOSIS

## DISEASES OF AORTA AND PULMONARY ARTERIES

| | Code Numbers |
|---|---|
| 1 Aneurysm of aorta (*specify location*) | 46 — 6 |
| 2 Aneurysm of pulmonary artery | 4711— 6 |
| 3 Aortitis | 461—1 |
| 4 Arteriosclerosis of aorta | 461—912 |
| 5 Arteriosclerosis of pulmonary arteries | 471—912 |
| Congenital anomaly of aorta or pulmonary artery (see diagnosis 38) | |
| 6 Dissecting hematoma of aorta (dissecting aneurysm) (*specify location*) | 46 —911 7<br>46 —911 6* |
| 7 Embolism of aorta | 4616—196 1 |
| 8 Embolism of pulmonary arteries (acute cor pulmonale) | 471—196 1 |
| 9 Injury of aorta or pulmonary artery | 461—4<br>4711—4 |
| 10 Other diseases of aorta (*specify lesion*) | 461— |
| 11 Other diseases of pulmonary arteries (*specify lesion*) | 471— |
| 12 Rupture of aorta spontaneous | 46 — 5 |
| 13 Thrombosis of aorta | 46 — 7 |
| 14 Thrombosis of pulmonary arteries | 471— 7 |

## DISEASES OF CORONARY ARTERIES

| | |
|---|---|
| 15 Arteriosclerosis of coronary arteries | 41x—912 |
| 16 Arteritis of coronary arteries | 41x—100 |
| Congenital anomaly of coronary artery (see diagnosis 38) | |
| 17 Embolism of coronary artery | 41x—196 1 |
| 18 Other diseases of coronary artery (*specify*) | 41x— |
| 19 Periarteritis nodosa of coronary arteries | 41x—931 0 |
| 20 Stenosis of coronary orifice | 41x— 4 |
| 21 Thrombosis of coronary artery | 41x— 7 |
| 22 Trauma of coronary artery (*specify character of lesion*) | 41x—4 |

## DISEASES OF ENDOCARDIUM AND VALVES

| | | Code Numbers |
|---|---|---|
| | Congenital anomaly of endocardium or valves (see diagnosis 38) | |
| 23 | Endocarditis acute bacterial (*specify organism*) | 150–1 |
| 24 | Endocarditis indeterminate | 150–911 1 |
| 25 | Endocarditis subacute bacterial (endocarditis lenta) (*specify organism*) | 150–1 |
| 26 | Mural endocarditis | 156–1 |
| 27 | Mural thrombosis | 156–1 7<br>–511 7<br>–932 7 |
| 28 | Neoplasm of endocardium | 150–8 |
| 29 | Other structural diseases of endocardium or valves (*specify location if possible*) | 150–<br>151– |
| 30 | Rupture of valve or of chordae tendineae (*specify valve*) | 151–1 5<br>157–1 5 |
| 31 | Traumatic injury of endocardium or valves (*specify lesion*) | 151–4<br>157–4 |
| 32 | Valvular deformity | |
| | a Aortic insufficiency | 455– x |
| | b Aortic stenosis | 155– 1 |
| | c Mitral insufficiency | 154– \ |
| | d Mitral stenosis | 154– 1 |
| | e Tricuspid insufficiency | 152– x |
| | f Tricuspid stenosis | 152– 1 |
| 33 | Valvular sclerosis (*specify valve affected*) | 15 –911 |
| 34 | Valvulitis active (*specify deformity, if any*) | 15 – |
| 35 | Valvulitis inactive (*specify deformity if any*) | 15 – 0 |

## DISEASES OF MYOCARDIUM
(Including Conduction System and Heart as a Whole)

| | | |
|---|---|---|
| 36 | Aneurysm of heart (*specify location*) | 11x–912 6 |
| 37 | Atrophy of heart | 110–701<br>–798<br>–911 |
| 38 | Congenital anomaly of heart or great vessels (*specify lesion if possible*) | 110–010 |

## A Non Cyanotic Group

| | | Code Numbers |
|---|---|---|
| A 1 | Aneurysm of sinus of Valsalva congenital | 4612–015 |
| A 2 | Aortic ring double aortic arch | 461–019 |
| A 3 | Aortic stenosis | 455–017 |
| A 4 | Bicuspid aortic valve | 455–0242 |
| A 5 | Coarctation of aorta | |
| | (1) Infantile type | 461–018 |
| | (2) Adult type | 461–0181 |
| A 6 | Dextrocardia | 410–021 |
| A 7 | Dilatation of pulmonary artery primary | 4711–015 |
| A 8 | Hypertrophy of heart congenital | 410–013 |
| A 9 | Hypoplasia of aorta | 461–016 |
| A10 | Left coronary artery arising from pulmonary artery | 41x2–010 |
| A11 | Pulmonary veins all drain into right atrium | 486–021 |
| A12 | Pulmonary veins entering right atrium or superior vena cava | 486–022 |
| A13 | Pulmonic stenosis isolated | 4711–018 |
| A14 | Right aortic arch | |
| | (1) Right aortic arch with aorta descending on right | 4613–019 |
| | (2) Right aortic arch with left descending aorta | 4613–0192 |
| A15 | Subaortic stenosis | 456–019 |
| A16 | Tricuspid valves in anomalous position | 452–021 |

## B Potentially Cyanotic Group
(cyanose tardive)

| | | |
|---|---|---|
| B 1 | Atrial septal defect | 412–0xx<br>–030<br>–0x9 |
| B 2 | Combined atrial and ventricular septal defects (persistent atrioventricularis communis) | 412–0x3 |
| B 3 | Patent ductus arteriosus | 40x–0x0 |
| B 4 | Patent foramen ovale (persistent ostium secundum) | 412–0x2 |
| B 5 | Ventricular septal defect (maladie de Roger) | 413–0x5 |

## DISEASES OF ENDOCARDIUM AND VALVES

| No. | Diagnosis | Code Numbers |
|---|---|---|
| | Congenital anomaly of endocardium or valves (see diagnosis 38) | |
| 23 | Endocarditis acute bacterial (*specify organism*) | 450–1 |
| 24 | Endocarditis indeterminate | 450–911 1 |
| 25 | Endocarditis subacute bacterial (endocarditis lenta) (*specify organism*) | 450–1 |
| 26 | Mural endocarditis | 456–1 |
| 27 | Mural thrombosis | 456–1 7<br>–511 7<br>–932 7 |
| 28 | Neoplasm of endocardium | 450–8 |
| 29 | Other structural diseases of endocardium or valves (*specify location if possible*) | 450–<br>451– |
| 30 | Rupture of valve or of chordae tendineae (*specify valve*) | 451–1 5<br>457–1 5 |
| 31 | Traumatic injury of endocardium or valves (*specify lesion*) | 451–1<br>457–1 |
| 32 | Valvular deformity | |
| | a Aortic insufficiency | 455– x |
| | b Aortic stenosis | 455– 1 |
| | c Mitral insufficiency | 451– x |
| | d Mitral stenosis | 451– 1 |
| | e Tricuspid insufficiency | 452– x |
| | f Tricuspid stenosis | 452– 1 |
| 33 | Valvular sclerosis (*specify valve affected*) | 45 –911 |
| 34 | Valvulitis active (*specify deformity if any*) | 45 – |
| 35 | Valvulitis inactive (*specify deformity if any*) | 45 – 0 |

## DISEASES OF MYOCARDIUM
(Including Conduction System and Heart as a Whole)

| No. | Diagnosis | Code Numbers |
|---|---|---|
| 36 | Aneurysm of heart (*specify location*) | 11x–912 6 |
| 37 | Atrophy of heart | 110–701<br>–799<br>–911 |
| 38 | Congenital anomaly of heart or great vessels (*specify lesion if possible*) | 110–010 |

| | Code Numbers |
|---|---|
| 47 Other structural diseases of heart (*specify lesion*) | 4 |
| 48 Rupture of myocardium (*specify location*) | 4 –4 .5 |
| | –416 |
| | –5x7 5 |
| 49 Thrombosis within heart (*specify chambers*) (See also Mural thrombosis) | 43 –522 7 |
| | –5x4 |
| 50 Trauma of myocardium | 43x–4 |
| 51 Undiagnosed structural disease of heart (*specify location if possible*) | 4 –9x0 |

## DISEASES OF PERICARDIUM

| | |
|---|---|
| 52 Calcification of pericardium | 420–923 |
| 53 Congenital anomaly of pericardium (*specify lesion*) | 420–0 |
| 54 Hemopericardium | 420–4 7 |
| 55 Hydropericardium | 420–522 8 |
| | –900 8 |
| 56 Neoplasm of pericardium | 420–8 |
| 57 Other structural diseases of pericardium | |
| 58 Pericarditis acute | 420–1 |
| a Fibrinous | 420–511 7 |
| | –932 |
| b Serofibrinous | 420–1 8 |
| c. Suppurative | 420–1 2 |
| 59 Pericarditis chronic | 420–100 0 |
| a Adhesive without constriction | 420–1x0 6 |
| b Constrictive | 420–1x0 1 |
| c Serous | 420–1x0 8 |
| 60 Pneumopericardium | 420–1 3 |
| | –4 3 |
| 61 Trauma to pericardium (*specify character of lesion*) | 420–4 |

# PHYSIOLOGICAL DIAGNOSIS

## CARDIAC MECHANISM

| | |
|---|---|
| 1 Arrhythmia (*undiagnosed*) | 451 |
| 2 Atrial Fibrillation | |
| a Transient | 425 |
| b Persistent | 426 |

## C Cyanotic Group

| | | Code Numbers |
|---|---|---|
| | C 1 Aortic atresia or stenosis | 455–017 |
| | C 2 Eisenmenger complex | 413–0x3 |
| | C 3 Persistent truncus arteriosus | 4051–01x |
| | C 4 Single ventricle with rudimentary outlet chamber (cor triloculare biatriatum) | 413–0x8 |
| | (1) When pulmonary artery arises from rudimentary chamber | |
| | (2) When great vessels are transposed aorta arising from rudimentary chamber pulmonary artery from common chamber | |
| | C 5 Tetralogy of Fallot | 413–0x0 |
| | C 6 Transposition of great vessels complete or partial | 4051–02x |
| | | –02x1 |
| | C 7 Tricuspid stenosis or atresia (pseudo triloculare) | 452–018 |
| 39 | Degeneration of myocardium (*specify variety if possible*) | 430–511 |
| | | –516 |
| | | –922 |
| | | –955 |
| 40 | Enlargement of heart (*chambers involved may be specified*) | |
| | a Dilatation | 410–13x |
| | b Hypertrophy | 410–13x 6 |
| 41 | Fatty infiltration of heart | 410–754 |
| 42 | Fibrosis of myocardium | 430–511 6 |
| | | –516 6 |
| | | –912 6 |
| | | –955 |
| 43 | Infarct of myocardium | 430–511 7 |
| | | –512 7 |
| | | –515 7 |
| | | –516 7 |
| | a Atrial infarction | 431–51 7 |
| | b Ventricular infarction | 431–51 7 |
| 44 | Myocarditis active | 430–930 |
| | | –932 |
| 45 | Neoplasm of heart (*specify type*) | 410–8 |
| 46 | No structural disease of heart | 410–000 |

## CLINICAL SYNDROMES

| | |
|---|---|
| 20 Adams Stokes Syndrome | 4ɔɹ |
| 21 Anginal Syndrome | 401 |
| 22 Cardiac insufficiency | 101 |
| 23 Carotid Sinus Syndrome | 9681~ɔ81x |

## FUNCTIONAL CAPACITY

| | |
|---|---|
| Class I | 4ɔ7 |
| Class II | 4ɔ8 |
| Class III | 4ɔ9 |
| Class IV | 4ɔx |

## THERAPEUTIC CLASSIFICATION

Class A
Class P
Class C
Class D
Class E

## NO HEART DISEASE 410–

## PREDISPOSING ETIOLOGICAL FACTOR

## UNDIAGNOSED MANIFESTATION 410–Y00

| | Code Numbers |
|---|---|
| 3 Atrial Flutter | |
| a Transient | 123 |
| b Persistent | 124 |
| 4 Atrioventricular block | |
| a Incomplete A V block (prolonged conduction time) | 134 |
| b Incomplete A V block | 435 |
| c Complete A V block | 136 |
| 5 Atrioventricular Nodal Rhythm | 433 |
| 6 Escaped beats (ventricular escape) | 113 |
| 7 Intraventricular block | 437 |
| 8 Other arrhythmias *(specify)* | |
| 9 Paroxysmal tachycardia | |
| a Atrial | 122 |
| b Atrioventricular nodal | 132 |
| c Unknown supraventricular origin | 160 |
| d Ventricular | 442 |
| 10 Premature contractions | |
| a Atrial | 121 |
| b Atrioventricular | 131 |
| c Unknown supraventricular origin | 156 |
| d Ventricular | 141 |
| 11 Pulsus alternans | 154 |
| 12 Sinus arrest (sino atrial block) | 411 |
| 13 Sinus arrhythmia | 112 |
| 14 Sinus bradycardia | 113 |
| 15 Sinus rhythm normal | 150 |
| 16 Sinus tachycardia | 114 |
| 17 Ventricular fibrillation | 145 |
| 18 Wandering pacemaker | 415 |

## VALVULAR INCOMPETENCE

| | |
|---|---|
| 19 Valvular Incompetence | |
| a Aortic incompetence | 408 |
| b Mitral incompetence | 407 |
| c Pulmonic incompetence | 406 |
| d Tricuspid incompetence | 405 |

CRITERIA FOR CARDIAC DIAGNOSIS

# CRITERIA FOR CARDIAC DIAGNOSIS

## ETIOLOGICAL DIAGNOSIS

A knowledge of the causes of heart disease helps to guide prognosis and treatment it often aids in the diagnosis of the anatomical or physiological disturbances The etiological diagnosis is to be derived from the history of the past and present illnesses from the age of the patient and from consideration of the character of the structural and functional disturbances Occasionally the history is unconvincing and the anatomical diagnosis is not definitely indicative of any one etiological factor If two or more possible causes of heart disease are discovered each one that might be responsible for the presenting disease should be mentioned

1 Anemia —It is doubtful that anemia alone can cause structural heart disease other than enlargement due to dilatation Anemia however can give rise to abnormal physical signs and to *physiological disturbances* The systolic murmur of mitral incompetence systolic murmurs at either the pulmonic or the aortic area and cardiac arrhythmia are the most frequent abnormal physical signs Sinus tachycardia and *cardiac* insufficiency associated with high cardiac output are the common physiological disturbances Anemia may precipitate the anginal syndrome in persons with coronary arteriosclerosis

*Criteria for Diagnosis of Heart Diseases Due to Anemia*

a The presence of significant anemia
b The disturbances of cardiac function mentioned above
c Disappearance of signs and symptoms following correction of the anemia

2 Animal Parasites —*Trichina* plasmodium malariae echinococus and Rickettsia may cause myocardial changes leading to temporary electrocardiographic abnormalities

Trypanosomiasis leads to chronic myocarditis and fibrosis so that electrocardiographic changes arrhythmias and cardiac insufficiency may occur in middle life

*Criteria for Diagnosis of Heart Disease Due to Animal Parasites*

a Demonstration of the presence of the parasite within the body
b Evidence of involvement of the heart

*Criteria for Diagnosis of Bacterial Infection of the Heart*

a Demonstration of pathogenic bacteria by culture from blood or pericardial fluid

b Signs or symptoms indicating involvement of the endocardium myocardium or pericardium

5 CONGENITAL ANOMALY —This diagnosis is based on the finding of characteristic physical signs fluoroscopic or roentgenographic evidence possibly combined with measurements of intracardiac pressure and oxygen content of the blood in various parts of the heart The details are found under the individual diagnosis in the Anatomical Section

6 HYPERTENSION —Hypertension is considered to be present when recorded by the recommended technique the systolic blood pressure is persistently above 140 mm Hg and the diastolic above 90 mm Hg Hypertension is not to be considered as the etiological factor in heart disease when the systolic phase alone is elevated as may be found in thyrotoxicosis heart block aortic insufficiency and marked arteriosclerosis of the aorta

Permanent hypertension may be associated with chronic renal disease such as diffuse glomerulo nephritis atrophic pyelonephritis or polycystic renal disease or unassociated with any demonstrable disease in the latter case it is known as essential hypertension Essential hypertension may or may not coexist with arteriosclerosis Acute and transient forms of hypertension such as accompany acute diffuse glomerulonephritis pheochromocytoma or the specific hypertensive disease of pregnancy may be the cause of cardiac disease

*Criteria for Diagnosis of Heart Disease Due to Hypertension*

Persistent hypertension associated with evidence of heart disease

7 HYPERTHYROIDISM Hyperthyroidism usually produces functional cardiovascular disturbances The most common of these are sinus tachycardia and paroxysmal or permanent atrial fibrillation or flutter Cardiac insufficiency may appear and usually with an increased cardiac output

A large adenoma may cause shortness of breath by pressure on the neighboring structures but disturbance of cardiac function and structural disease of the heart are not produced in such cases unless disease of the thyroid gland is accompanied by hyperthyroidism

3 ARTERIOSCLEROSIS — For the purposes of this classification the etiological diagnosis Arteriosclerosis should be limited to those cases of heart disease showing definite evidence of arteriosclerosis of the coronary arteries or the arch of the aorta

The secondary structural changes in the myocardium result from inadequate coronary blood supply and consist of degeneration and fibrosis The extent of such changes varies widely in different patients and is not necessarily proportional to the symptoms of which the patient complains Although the main lesions of arteriosclerotic heart disease are found in the coronary arteries and the myocardium the aorta and the heart valves may likewise be involved Occasionally the latter are sufficiently affected to produce insufficiency of the aortic valve or stenosis of its orifice

Cardiac insufficiency or the anginal syndrome may result when the coronary blood flow is inadequate Atrial fibrillation and premature beats often occur or there may be evidences of defective atrio ventricular or intraventricular conduction Coronary artery stenosis thrombosis or occlusion are common Enlargement of the heart usually is not present unless there is either associated hypertension congestive heart failure persistent arrhythmia or myocardial infarction When arterial hypertension is also present it should be recorded as an additional etiological diagnosis

*Criteria for Diagnosis of Arteriosclerotic Heart Disease*

The diagnosis rests with the finding of symptoms or abnormal physical signs indicating

a Arteriosclerosis of the coronary arteries
b Thrombosis or occlusion of one or more coronary branches
c Fibrosis of the myocardium
d Sclerosis of a valve
e Arteriosclerosis of the aorta

4 BACTERIAL INFECTION — The endocardium myocardium and pericardium are each subject to bacterial infection This diagnosis is to include infection by bacteria such as Streptococcus viridans Streptococcus hemolyticus Staphylococcus aureus or albus Bacillus influenzae Pneumococcus Gonococcus and Meningococcus or others The tubercle bacillus rarely invades the endocardium or myocardium but not infrequently involves the pericardium

The diagnostic criteria depend upon the demonstration of signs and symptoms of involvement of the endocardium the myocardium or the pericardium by one of the organisms mentioned above

*Criteria for Diagnosis of Bacterial Infection of the Heart*

a Demonstration of pathogenic bacteria by culture from blood or pericardial fluid

b Signs or symptoms indicating involvement of the endocardium myocardium or pericardium

5 CONGENITAL ANOMALY – This diagnosis is based on the finding of characteristic physical signs fluoroscopic or roentgenographic evidence possibly combined with measurements of intracardiac pressure and oxygen content of the blood in various parts of the heart The details are found under the individual diagnosis in the Anatomical Section

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Permanent hypertension may be associated with chronic renal disease such as diffuse glomerulo-nephritis atrophic pyelonephritis or polycystic renal disease or unassociated with any demonstrable disease in the latter case it is known as essential hypertension Essential hypertension may or may not coexist with arteriosclerosis Acute and transient forms of hypertension such as accompany acute diffuse glomerulo nephritis pheochromocytoma or the specific hypertensive disease of pregnancy may be the cause of cardiac disease

*Criteria for Diagnosis of Heart Disease Due to Hypertension*

Persistent hypertension associated with evidence of heart disease

7 HYPERTHYROIDISM – Hyperthyroidism usually produces functional cardiovascular disturbances The most common of these are sinus tachycardia and paroxysmal or permanent atrial fibrillation or flutter Cardiac insufficiency may appear and usually with an increased cardiac output

A large adenoma may cause shortness of breath by pressure on the neighboring structures but disturbance of cardiac function and structural disease of the heart are not produced in such cases unless disease of the thyroid gland is accompanied by hyperthyroidism

*Criteria for Diagnosis of Heart Disease Due to Hyperthyroidism*

Evidence of hyperthyroidism associated with abnormal cardiac function such as sinus tachycardia premature beats atrial fibrillation or flutter or cardiac insufficiency or with cardiac enlargement

8 HYPOTHYROIDISM —The lack of adequate thyroid secretion has certain effects upon the heart There may be cardiac enlargement changes in the electrocardiogram especially a prolonged P R interval low voltage QRS and abnormal or inverted T waves There may at times be signs of cardiac insufficiency If diminution in the size of the heart and reversion of the electrocardiographic abnormalities result from the administration of desiccated thyroid the etiological relationship is established When hypothyroidism occurs in patients with arteriosclerosis both etiological factors should be entered in the diagnosis

*Criteria for Diagnosis of Heart Disease Due to Hypothyroidism*

a Evidence of hypothyroidism
b *Enlargement of the heart electrocardiographic changes and possibly cardiac insufficiency*
c Favorable response to the administration of thyroid extract

9 *NEOPLASM —Primary new growths in the heart are rare* They may be suspected in patients with otherwise unexplained signs and symptoms such as compression or thrombosis of the superior vena cava pericardial effusion intermittent and varying types of arrhythmias especially atrial fibrillation and cardiac insufficiency Diagnostic certainty is achieved by finding tumor cells in the pericardial exudate Diagnostic pneumopericardium may be of value as a part of the x ray study

*Secondary* tumors of the heart are either metastatic or produced by extension of intrathoracic tumors chiefly bronchial The atria are usually the first portion of the heart to be invaded Secondary involvement of the heart by tumor is more likely to be recognized than is primary involvement

10 NEUROCIRCULATORY ASTHENIA (effort syndrome) —Manifestations are unusual fatigability pallor or flushing tremor cyanosis of the extremities axillary and palmar sweating palpitation dyspnea faintness and precordial pain These symptoms may be present at rest but are characteristically induced by physical effort or emotional disturbances and are excessive in proportion to the amount of effort or to the

strength of the emotional stimulu They are most frequently found in young males of asthenic habitus The predominance of evidence indicates that this condition develops on the basis of a neurosis When neurocirculatory asthenia is the only etiological diagnosis the anatomical diagnosis should be no structural disease

11 OTHER ETIOLOGICAL FACTORS (to be specified)—Heart disease may be associated with chronic nephritis diabetes obesity or gout In such cases arteriosclerosis or hypertension or both may be present and if so one or both of these should be recorded as the primary etiological diagnosis The general disease being a secondary diagnosis should be listed as an Accompanying condition

Parasites have been known to invade the heart and cause structural damage

In all cases of heart disease in which any uncertainty exists as to cause the etiological diagnosis should be stated as Unknown and the questionable etiological factor entered as an Accompanying condition

12 OTHER INFECTIONS—Any acute infection may aggravate existing heart disease but unless there is conclusive evidence that the infection has caused the heart disease the primary etiological diagnosis should be stated and the acute infection added as an Accompanying Condition There is evidence that viral infection may produce acute pericarditis and also that in certain other infections permanent cardiac damage may occur e g poliomyelitis

13 PULMONARY ARTERIAL HYPERTENSION—Increased pulmonary arterial pressure which is secondary to increased pressure in the pulmonary veins is not to be included under this title

An abnormally high pressure in the pulmonary arteries may occur acutely as with pulmonary embolism or may be chronic The chronic form is most frequently due to disease of the pulmonary parenchyma such as emphysema or pulmonary fibrosis These parenchymal changes also may result from certain anatomical changes in the bony thorax such as kyphoscoliosis or thoracoplasty Pulmonary arterial hypertension also may be due to changes in the pulmonary arterial system such as arteriolar sclerosis or to extrinsic pressure upon the pulmonary artery The right ventricle enlarges and eventually fails

Pulmonary arterial hypertension can be recognized with certainty only by measurement of pressure within the pulmonary artery by the use of a catheter It may be suspected when there is chronic pulmonary

disease or thoracic deformity associated with a loud pulmonic second sound or when there is roentgenologic demonstration of dilated pulmonary arteries or of enlargement of the right ventricle and atrium There also may be electrocardiographic evidence suggesting hypertrophy of these chambers

The symptoms due to the pulmonary disease become aggravated when the right ventricle fails Signs and symptoms of systemic venous congestion appear and cyanosis is apt to be marked

*Criteria for the Diagnosis of Heart Disease Due to Pulmonary Arterial Hypertension*

The presence of one or more of the causes of pulmonary arterial hypertension associated with

a Pulmonary arterial hypertension either measured or suspected because of an accentuated pulmonic second sound or dilatation of pulmonary arteries

b Evidence of enlargement or insufficiency of the right ventricle and atrium

14 REFLEX DISTURBANCE OF THE HEART —Certain patients may be subject to physiological disturbances of the heart action as a result of reflexes arising in other parts of the body such as the carotid sinus or the abdominal or thoracic viscera Premature contractions are the most frequent of such disturbances but paroxysmal tachycardia atrial fibrillation sinus bradycardia, cardiac standstill or heart block also may occur

*Criteria for Diagnosis of Reflex Disturbance of the Heart*

a Presence of cardiac arrhythmia and of disease or disturbed function of some structure other than the heart

15 RHEUMATIC FEVER —Rheumatic fever is a generalized disease of the connective tissue with the clinical and epidemiological features of an infectious disease Group A hemolytic streptococcus plays a dominant role in the etiology Both the initial attack and the recurrences usually follow in the wake of an upper respiratory infection due to these organisms or of scarlet fever

The symptoms and course of rheumatic fever are extremely variable Three major clinical manifestations are migratory polyarthritis Sydenham s chorea and carditis *The term carditis is used to denote acute* pericarditis myocarditis or endocarditis separately or in combination

Especially in warm climates carditis is prone to occur without there having been chorea or polyarthritis Certain minor manifestations such as muscle or joint pains epistaxis erythema multiforme subcutaneous nodules anorexia abdominal pain low-grade fever abnormal fatigability and retardation of growth should be considered as possibly due to rheumatic fever The tendency to rheumatic relapses is most marked during childhood and declines after puberty The likelihood of cardiac involvement increases with each recurrence

Whenever a diagnosis of rheumatic heart disease is made the physician should state whether the rheumatic process is active or *inactive* Patients with low grade fever persistent tachycardia nodules joint involvement choreiform movements erythema elevated sedimentation rate leucocytosis or changing electrocardiograms should be classified as active provided that there is no other discoverable cause for these findings If these features are not present the case should be classified as inactive

*Criteria for the Diagnosis of Rheumatic Heart Disease*

a History of polyarthritis chorea or the above mentioned minor manifestations accompanied by a characteristic structural lesion of the heart

b Evidence of a characteristic structural lesion of the heart even without a history of rheumatic fever or of the above mentioned minor manifestations

16 SYPHILIS —Heart disease may be produced by the activity of the *Treponema pallidum* in the wall of the *aorta* The *myocardium* is commonly affected as a result of syphilitic aortitis *involving the orifices* of the coronary arteries Gumma of the *myocardium* is rare The predominant lesion is in the ascending aorta *frequently complicated* by one or more of the following *Aortic valve deformity with regurgitation* dilatation or aneurysm *of the aorta and coronary* ostial stenosis Syphilitic aortitis usually is present for a *considerable number* of years before symptoms of heart disease *appear It may* therefore be possible to *recognize it before* the appearance of symptoms by the discovery of aortitis or of aortic insufficiency

Though signs of aortic involvement may be discovered much earlier the average time elapsing *from the date of the original* infection until the *appearance of symptoms* of heart disease is from fifteen to twenty years The duration of life *after the* appearance of symptoms is much shorter

disease or thoracic deformity associated with a loud pulmonic second sound or when there is roentgenologic demonstration of dilated pulmonary arteries or of enlargement of the right ventricle and atrium There also may be electrocardiographic evidence suggesting hypertrophy of these chambers

The symptoms due to the pulmonary disease become aggravated when the right ventricle fails Signs and symptoms of systemic venous congestion appear and cyanosis is apt to be marked

*Criteria for the Diagnosis of Heart Disease Due to Pulmonary Arterial Hypertension*

The presence of one or more of the causes of pulmonary arterial hypertension associated with

a Pulmonary arterial hypertension either measured or suspected because of an accentuated pulmonic second sound or dilatation of pulmonary arteries

b *Evidence of enlargement or insufficiency of the right ventricle and atrium*

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The symptoms and course of rheumatic fever are extremely variable Three major clinical manifestations are migratory polyarthritis Sydenham s chorea and carditis The term carditis is used to denote acute pericarditis myocarditis or endocarditis separately or in combination

ruptured valvular cusps or rupture of the aorta but only if there is pre existing disease of these structures

*Criteria for Diagnosis of Heart Disease Due to Trauma*

a The existence of adequate trauma as described

b Evidence of cardiac disease

20 UNKNOWN—The etiology should be stated as Unknown in those cases which present either definite structural changes in the heart or evidence of abnormal cardiac function for which no definite etiology can be determined

A certain number of the cases of unknown etiology are probably due to rheumatic fever the heart disease being its only manifestation Such cases in the absence of a definite rheumatic history may present the lesions typical of rheumatic heart disease and are of the age at which this is commonly found Similarly a certain number may be due to syphilis though the syphilitic infection cannot be proved clinically

*Criteria for Diagnosis of Unknown Etiology*

The absence of a definite etiology in the presence of

a Structural lesion of the heart

b Abnormal cardiac function

*Criteria for Diagnosis of Syphilitic Heart Disease*

a A history of syphilitic infection with evidence of a characteristic structural lesion of the aorta or aortic valve

b A characteristic structural lesion of the aorta or of the aortic valve without a history of syphilis but with a positive serological test

*c A characteristic structural lesion of the aorta or of the aortic valve* together with evidences of syphilitic disease elsewhere such as cerebrospinal syphilis even in the absence of a positive serological test for syphilis or a history of syphilitic infection

17 THIAMIN DEFICIENCY —This condition often occurs as a complication of chronic alcoholism and is common in beri beri One finds general cardiac enlargement and abnormal features in the electrocardiogram most commonly a decrease in the size of the T waves Heart failure may occur in the later stages and is associated with a high cardiac output Diagnosis is confirmed if all signs and symptoms are relieved following the administration of thiamin chloride in adequate doses

18 TOXIC AGENT —This etiological diagnosis is to include those instances of abnormal cardiac function or structural cardiac disease clearly due to bacterial toxins or chemical substances These toxic agents should be specified A typical example of a bacterial toxin is that of the diphtheria bacillus Some commonly encountered chemical substances which may produce toxic effects are digitalis phosphorus quinidine and nicotine

This etiological diagnosis should not be made unless the particular agent is definitely identified If not established the etiology should be entered as unknown

*Criteria for Diagnosis of Heart Disease Due to Toxic Agent*

Presence of a definite toxic agent associated with

a Abnormal cardiac function

b Structural lesion of the heart

19 TRAUMA —Structural cardiac lesions and abnormal cardiac function occur as the result of penetrating wounds severe blows upon the chest or crushing injuries of the chest There is no conclusive evidence that either severe or prolonged and sustained physical effort can cause coronary artery thrombosis In the presence of coronary arteriosclerosis they may cause *coronary insufficiency* with myocardial ischemia perhaps leading to cardiac infarction Severe physical effort may produce

of abnormal areas of marked dullness The accuracy of percussion as an indication of heart size leaves much to be desired especially in obese or emphysematous patients Therefore for determining the size and shape of the heart and great vessels roentgenologic methods should be employed whenever possible

*Auscultation* enables the examiner to recognize the first and second heart sounds to describe their characteristics and to detect and describe murmurs Heart sounds and murmurs are described with reference to the time of occurrence in the cardiac cycle their intensity quality and duration and the site of maximum intensity For determining the time of occurrence of murmurs in the cardiac cycle it is necessary first to identify the first and second heart sounds This may be effected by simultaneous palpation of the apex impulse or the carotid pulse In the case of murmurs it is useful to note the area of maximum audibility the direction of their transmission and their modification by respiration posture and exercise Changes in the quality of sounds and murmurs observed on successive examinations should be noted

Identical sounds and murmurs are frequently described in *different* words by different observers This is due in part to differences in the ability of individuals to classify auditory impressions in part to the *multiplicity of terms* in general usage Many of these terms are synonymous and it is believed that an attempt to standardize descriptive terms will prove useful The following descriptive terms all in common use will serve to characterize most heart sounds and murmurs If in exceptional cases these terms seem inadequate it is suggested that the most nearly satisfactory ones from this list be used together with such additional words as may be deemed necessary In the table the terms in capitals (FAINT) are recommended for *routine* use Those in smaller type (weak) are regarded as less satisfactory synonyms for those terms in capitals immediately *preceding* them but may be used as additions if it is thought that the recommended term is *inadequate* The following terms are *suggested* for the various gradations of intensity Very faint, faint moderate loud very loud

*Cardiorespiratory murmurs* are sometimes confusing in that they may simulate those indicating cardiac disease They are usually heard at the apex or over the body of the heart at the lung margins They are almost always systolic in time and vary in intensity during the phases of respiration Holding the breath during a forced expiration sometimes diminishes or abolishes them

*Innocent murmurs* —Certain murmurs occur in the absence of structural changes in the heart They have often been called functional accidental or hemic The use of these terms is to be discouraged

# ANATOMICAL DIAGNOSIS

A CORRECT anatomical diagnosis can be made clinically in the majority of the diseases of the heart The various diagnostic methods that should be employed are (1) Complete history and physical examination (2) electrocardiography (3) roentgenology including fluoroscopy and where indicated angiocardiography (4) Other laboratory tests are recommended Serological tests for syphilis urine examination blood count and erythrocyte sedimentation rate non protein nitrogen and *other renal function tests when indicated*

To compile a complete catalogue of the signs of the structural changes associated with the different diseases of the heart is beyond the scope of this chapter An attempt has been made to set down those features regarded as of major diagnostic importance together with a few such minor criteria as may on occasion prove helpful

In making the examination it is best to follow a routine including inspection palpation percussion and auscultation

*Inspection* should include a search for the presence of dyspnea cyanosis pallor jaundice petechiae subcutaneous nodules rashes clubbing of the fingers pulsations or engorgement of the veins and chest deformities in the heart region a search for the apical thrust and other localized impulses or areas of retraction finally for any abnormal pulsations in the anterior and posterior walls of the chest and in the abdominal wall especially in the region of the liver

*Palpation* should be used to locate the apical impulse and to deter*mine its force and extent The point furthest downward and outward* at which a distinct thrust can be felt serves to indicate the position of the left border of the heart This point is usually a more useful guide to the left border than is percussion particularly when the heart is enlarged The site of maximum impulse usually lies mesial to this point Abnormal impulses and retractions thrills enlargement and pulsations of the liver and the presence of edema are also elicited by palpation The pulse its rate and character and the consistency of the arterial wall should be determined Palpation of the pulse may be mis leading in the determination of arrhythmias

*Percussion* serves to outline areas of marked dullness attributable to the heart and great vessels *Enlargement of the heart dilatation of the* great vessels and pericardial effusion may be suggested by the presence

Innocent murmurs are usually systolic in time more often faint than of moderate intensity and usually blowing in quality They are often inconstant and vary in intensity with change in posture

The commonest is a systolic murmur at the pulmonic area which is usually best heard with the patient supine and during expiration An apical systolic murmur is also common Such murmurs are unaccompanied by any evidence of structural disease such as an enlargement of the heart abnormal cardiac silhouette on roentgenologic examination or an abnormal electrocardiogram and do not indicate heart disease The mode of their production is unknown We have adopted the term *innocent murmur** to be applied to them considering it preferable to the title *unknown* which was previously recommended

## DISEASES OF AORTA AND PULMONARY ARTERIES

I ANEURYSM OF AORTA (specify location)—The term aneurysm should be limited to a saccular or sharply demarcated fusiform dilatation of the aorta In the thoracic aorta aneurysms are usually due to syphilis whereas in the abdominal aorta arteriosclerosis is the most common cause Aneurysms due to acute infections are rare and usually too small to be diagnosed before death Traumatic aneurysms of the aorta are uncommon Aneurysms of the aorta may remain latent without signs or symptoms When present these are due to pressure on or erosion of adjacent structures Pressure on a bronchus not infrequently results in bronchiectasis

The so called dissecting aneurysm of the aorta is discussed under the heading Dissecting Hematoma of Aorta

*Symptoms and Signs*

- Cough hoarseness and dysphagia
- Localized thoracic pain
- Inequality of the pulse and blood pressure in the two arms
- Tracheal tug
- Localized pulsation at the point where the aneurysm impinges upon the chest wall
- Abnormal dullness over the manubrium sterni or in the first and second interspaces to the right or left of the sternum
- Recurrent laryngeal or phrenic nerve paralysis
- Fluoroscopy may show increased localized pulsation of the vessel
  Roentgenography may reveal sacculation or a sharply demarcated fusiform dilatation of some parts of the aorta

* This term has been used by William Evans *Brit Heart J* 19[illegible]

## TABLE OF TERMS TO BE USED IN DESCRIBING HEART SOUNDS AND MURMURS

### HEART SOUNDS

| Intensity | Quality | Duration |
|---|---|---|
| NORMAL | NORMAL | NORMAL |
| VERY FAINT | SNAPPING<br>sharp<br>valvular | SHORT |
| FAINT<br>weak<br>muffled<br>distant | BOOMING<br>muscular | PROLONGED |
| LOUD<br>accentuated<br>increased | SPLIT<br>REDUPLICATED | |
| VERY LOUD | RINGING<br>metallic<br>bell like<br>tambour<br>hollow | |
| ABSENT<br>replaced by a murmur | | |

### MURMURS

| Intensity | Pitch | Quality | Duration | Time |
|---|---|---|---|---|
| VERY FAINT | HIGH | BLOWING | SHORT | SYSTOLIC |
| FAINT<br>soft | LOW | HARSH<br>rough<br>coarse | MODERATE | EARLY SYSTOLIC |
| | | | LONG | LATE SYSTOLIC |
| MODERATE | | MUSICAL<br>clicking | | DIASTOLIC |
| LOUD | | RUMBLING | | EARLY DIASTOLIC<br>protodiastolic |
| VERY LOUD | | CRESCENDO | | MID DIASTOLIC |
| | | DECRESCENDO<br>diminuendo | | PRESYSTOLIC<br>late diastolic |

*Symptoms*

None

*Signs*

Ringing quality of the aortic second sound with or without increased intensity

A systolic murmur heard loudest at the aortic area

Increase in width of the aortic silhouette on fluoroscope radiography orthodiagraphy or angiocardiography

Increased amplitude of pulsation in the ascending transverse or descending thoracic aorta demonstrated on fluoroscopy or by electrokymography

A localized increased convexity may be seen on the lateral or ventral aspect of the ascending aorta

Calcification of the ascending aorta occasionally may be observed

4 ARTERIOSCLEROSIS OF AORTA —This is usually minimal in the ascending portion moderately severe in the arch and descending thoracic aorta and is generally most severe in the abdominal segment The condition may be found in young subjects but makes its appearance to a significant degree after age 40

*Signs*

Systolic murmur heard best over the second right interspace near the sternum.

Elongation uncoiling and increase in width and density of the aortic silhouette on fluoroscopy or radiography The descending aorta becomes readily visible and often curves well to the left Calcareous plaques may be demonstrated roentgenologically in the aortic knob and in the abdominal aorta (See Guide to Roentgenological Diagnosis)

5 ARTERIOSCLEROSIS OF PULMONARY ARTERIES is usually found in conditions associated with elevated pulmonary arterial pressure *e g* mitral stenosis pulmonary emphysema silicosis deformities of the thorax and congenital anomalies of the heart and great vessels It may occur in the absence of any of these lesions

The diagnosis is rarely made clinically because signs and symptoms are frequently lacking and when present are the result of the primary disease and its effects upon the right heart These will be described under the sections on acute and chronic pulmonary heart disease

Angiocardiography or electrokymography may help to differentiate an aneurysm from other tumors

The more sensitive serologic tests will be positive in a large percentage of cases when the aneurysm is due to syphilis

2 ANEURYSM OF PULMONARY ARTERY may be encountered in hearts with congenital malformations associated with marked pulmonary hypertension On rare occasions it is due to syphilis It usually can be diagnosed only by roentgenography

3 *AORTITIS usually is caused by syphilis more rarely by rheumatic fever and other infectious diseases but the form due to syphilis is the only one in which a clinical diagnosis can be made with reasonable certainty* The diagnosis of uncomplicated syphilitic aortitis is especially *difficult* in patients over 40 years of age or in those with hypertension arteriosclerosis or any other lesions that may lead to dilation *of the aorta*

*Syphilitic* aortitis frequently escapes clinical recognition since it never produces symptoms The appearance of symptoms indicates that *a complication* has arisen such as occlusion of one of the coronary ostia or aneurysm of the aorta Evidences of aortitis may be found within 5 to *10 years* after the chancre The blood Wassermann reaction is often negative but more sensitive serologic tests may *be expected to give a* much higher percentage of positive reactions In patients with syphilis under 40 years of age and whose blood pressure is *normal the diagnosis* may be suspected from the presence of a ringing *aortic second* sound more rarely from a systolic murmur over the aortic area

Although the demonstration of a widened aortic silhouette by radio*logical technics is one of the most important aids in the diagnosis of* uncomplicated syphilitic aortitis its appearance may be simulated by the effects of hypertension or of arteriosclerosis of the aorta A bulging of the ascending aorta in an individual of 40 years or less may *indicate* syphilitic aortitis particularly if this portion shows exaggerated pulsation under the fluoroscope

Syphilitic aortitis *is frequently complicated* by coronary *ostial* stenosis aortic insufficiency and aneurysm The presence of one or more of these lesions facilitates the recognition of the underlying aortitis

In patients with syphilis who are over age 40 and in whom dilatation of the aorta has been demonstrated by the criteria listed below a clinical diagnosis of Aortitis or Arteriosclerosis of the aorta should be made In this age group it is not possible to state with *certainty whether* syphilis or arteriosclerosis or a combination of the two lesions has caused the aortic dilatation

7 EMBOLISM OF AORTA —Emboli usually lodge at the bifurcation of the aorta Large emboli usually come from the heart In rheumatic heart disease they arise from thrombi in the left auricle In arteriosclerotic heart disease they are derived from mural thrombi in the left ventricle after myocardial infarction or more rarely from thrombi in the fibrillating left atrium or its auricle

It may be difficult and at times impossible to distinguish between thrombosis and embolism as the cause of sudden occlusion of the aorta However in patients with rheumatic heart disease or recent myocardial infarction embolism is more probable

*Symptoms*

Sudden onset of severe pain in the legs lower abdomen or back
In rare cases pain may be absent
Sudden onset of numbness coldness and tingling of both legs though usually one leg is more affected than the other

*Signs*

Pallor or mottled cyanosis of one or both legs
Coldness of one or both legs
Absence of arterial pulsation in all arteries of one or both lower extremities
Collapsed superficial veins of the extremity
Loss of power in one or both legs

8 EMBOLISM OF PULMONARY ARTERIES (acute cor pulmonale) — The common sites of thrombi which may give rise to pulmonary emboli are in the order of their frequency the deep veins of the feet and legs the femoral iliac pelvic and prostatic veins the right atrium or its auricle and the right ventricle

Symptoms result from infarction of the lung and vary in degree with the size of the vessel occluded If it is small there may be no symptoms The presence of embolism of a small pulmonary artery may be suspected when a rise in respiratory rate often initiated by chest oppression is followed by fever A large embolus may in addition cause sweating weakness collapse or sudden death When pain in the chest occurs it may be aggravated by deep breathing It may be retrosternal or may radiate to the left shoulder and thus closely resemble the pain of acute myocardial infarction Hemoptysis may occur soon after the onset of pain but more commonly it is delayed for several hours or may not occur at all

Congenital Anomaly of Aorta or Pulmonary Artery.—See Congenital Anomaly of Heart or Great Vessels.

6. Dissecting Hematoma of Aorta (*Dissecting Aneurysm*).—This condition results from rupture of the aorta through the intima into the media. Medial necrosis is usually the underlying lesion. Rarely the rupture is through an atheromatous ulcer. Hypertension often plays a part in the etiology. The initial rupture usually occurs in the ascending aorta or arch and more rarely in the descending portion. The hematoma may extend proximally and distally between the layers of the media as well as circumferentially. As it does so, it may occlude small arteries and compress the orifice of larger ones as they leave the aorta. *Death may ensue rapidly or after many hours and may result from* hemorrhage into neighboring organs or occlusion of important vessels, especially the coronary arteries. Spontaneous recovery may rarely occur. Second or even third attacks may follow, ending in fatal hemorrhage. Dissecting hematoma occurs predominantly in males and in the middle or older age groups.

*Symptoms and Signs*

Sudden onset of severe, tearing, or crushing pain of long duration and situated in the front or back of the chest, less often in the abdomen. The pain is at its maximum at the onset and may be accompanied by extreme prostration, dyspnea, and loss of consciousness.

Symptoms result from the pressure of the hematoma upon the branches of the aorta, causing dysfunction of the structures supplied by these vessels.

There may be hematuria or anuria if the renal vessels are affected.

There may be a picture suggesting an acute surgical abdominal condition if the mesenteric arteries are involved. Embolism of iliac arteries may be suggested if these vessels are involved.

Hypertension is often present.

Enlargement of the heart.

The appearance of a diastolic murmur at the base of the heart.

*Maintenance of high blood pressure until shock develops.*

Fever and leukocytosis.

Electrocardiogram is variable and not characteristic.

Roentgenographic evidence of widening of the aorta may be obtained in certain cases. This can best be demonstrated by comparison of serial films* or with films taken before the attack.

* Roentgenographic studies are inadvisable in the acute phase or early stages.

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It may be difficult and at times impossible to distinguish between thrombosis and embolism as the cause of sudden occlusion of the aorta However in patients with rheumatic heart disease or recent myocardial infarction embolism is more probable

*Symptoms*

Sudden onset of severe pain in the legs lower abdomen or back

In rare cases pain may be absent

Sudden onset of numbness coldness and tingling of both legs though usually one leg is more affected than the other

*Signs*

Pallor or mottled cyanosis of one or both legs

Coldness of one or both legs

Absence of arterial pulsation in all arteries of one or both lower extremities

Collapsed superficial veins of the extremity

Loss of power in one or both legs

8 EMBOLISM OF PULMONARY ARTERIES (acute cor pulmonale) —The common sites of thrombi which may give rise to pulmonary emboli are in the order of their frequency the *deep veins of the feet and leg* the femoral iliac pelvic and prostatic veins the right atrium or its auricle and the right ventricle

Symptoms result from infarction of the lung and vary in degree with the size of the vessel occluded If it is small there may be no symptoms The presence of embolism of a small pulmonary artery may be suspected when a rise in respiratory rate often initiated by chest oppression is followed by fever A large *embolus* may in addition cause sweating weakness collapse or sudden *death* When pain in the chest occurs it may be aggravated by deep breathing It may be retrosternal or may radiate to the left shoulder and thus closely resemble the pain of acute myocardial infarction *Hemoptysis* may occur soon after the onset of pain but more commonly it is delayed for several hours or may not occur at all

*Signs*

Rapid shallow respirations
Cyanosis
Fever pallor sweating and syncope
Dilatation of the neck veins
Hemoptysis may occur
Jaundice
Rapid weak pulse
Accentuation of the pulmonic second sound
Pleural friction rub and evidence of pulmonary consolidation
Electrocardiographic abnormalities are influenced by the extent of the pulmonary artery occlusion and by the nature of the electro cardiogram before the occlusion occurred At times a curve resembling that of right bundle branch block appears early differing from the usual complex in that the S T segment is depressed in Leads I II and in the left precordial leads and elevated in Lead $V_R$ and in the right precordial leads The defect in conduction usually disappears in 24 hours and the S T segment displacement soon after The features that remain are those most commonly seen A prominent $S_1$ and $Q_3$ with inverted T waves in Leads II III and in the right precordial leads Often the abnormalities of the curve are non specific in type involving the T waves only or there may be no deviation from normal
Roentgenographic examination at the bedside when possible may show areas of increased pulmonary density

9 INJURY OF AORTA OR PULMONARY ARTERY —Trauma due to compression stab or bullet wounds or the swallowing of foreign bodies may injure the aorta or pulmonary artery Such injuries are usually rapidly fatal Very rarely they may result in slow bleeding The diagnosis depends upon the presence of a history of trauma or evidence of the same

The aorta may rupture as the result of crushing injuries to the chest Bleeding usually occurs into the pericardial sac pleural space or mediastinum

10 OTHER DISEASES OF AORTA (specify lesion) —Under this diagnosis may be listed such diseases as are not included in the other categories The various forms of infectious aortitis not due to syphilis may be mentioned here including those occurring in septicemia and in the bacteremias with or without endocarditis and in rheumatic fever Here also may be included medial (senile) and selective (metastatic) calci

fication of the aorta each of which can be differentiated from the usually encountered intimal calcification of arteriosclerosis by necropsy only In this group should also be included medial necrosis This diagnosis is made at necropsy but may be suspected when a dissecting hematoma occurs

11 OTHER DISEASES OF PULMONARY ARTERIES (specify lesion) —Under this diagnosis may be filed diseases not previously mentioned such as arteritis due to rheumatic fever tuberculosis periarteritis nodosa septic infections parasitic invasions or syphilis These diagnoses are rarely made except at necropsy

12 RUPTURE OF AORTA (spontaneous) —This may be partial or complete It never occurs in a structurally intact aorta Medial necrosis degenerative and inflammatory processes are the most important etiological lesions especially when associated with arterial hypertension Coarctation of the aorta and hypoplasia more rarely predispose to rupture A syphilitic aorta very rarely ruptures unless an aneurysm is present Complete rupture may be rapidly fatal but with slow bleeding into the esophagus pericardium or pleural space death may be postponed for hours or days

13 THROMBOSIS OF AORTA —Mural thrombi of infectious nature usually occur in the ascending aorta They may be associated with bacterial endocarditis of the aortic valve Mural thrombi rarely produce symptoms Thrombosis with gradual occlusion is encountered most frequently in the abdominal portion near its bifurcation The thrombus usually forms on an arteriosclerotic plaque or atheromatous ulcer rarely on a syphilitic lesion At times a thrombus may occlude so abruptly that the symptoms may closely resemble those of Embolism of Aorta Partial and even complete occlusion of the lower portion of the abdominal aorta by thrombus has been found at necropsy without there being any symptoms indicating its presence This is probably because of the gradual development of adequate collateral circulation

Signs

Pulsations in the femoral arteries are diminished or absent
Evidence of deficient arterial blood flow in both feet

14 THROMBOSIS OF PULMONARY ARTERIES —Conditions such as congestive heart failure which are accompanied by pulmonary stasis may cause thrombosis in the pulmonary arteries Occasionally the thrombus

*Signs*

Rapid shallow respirations

Cyanosis

Fever pallor sweating and syncope

Dilatation of the neck veins

*Hemoptysis may occur*

Jaundice

Rapid weak pulse

*Accentuation of the pulmonic second sound*

Pleural friction rub and evidence of pulmonary consolidation

Electrocardiographic abnormalities are influenced by the extent of the pulmonary artery occlusion and by the nature of the electrocardiogram before the occlusion occurred At times a curve resembling that of right bundle branch block appears early differing from the usual complex in that the S T segment is depressed in Leads I II and in the left precordial leads and elevated in Lead $V_R$ and in the right precordial leads The defect in conduction usually disappears in 24 hours and the S T segment displacement soon after The features that remain are those most commonly seen A prominent $S_1$ and $Q_3$ with inverted T waves in Leads II III and in the right precordial leads Often the abnormalities of the curve are non specific in type involving the T waves only or *there may be no deviation from normal*

Roentgenographic examination at the bedside when possible may show areas of increased pulmonary density

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other rarer conditions It is always a part of a general disease The rheumatic form is an acute inflammatory reaction involving the arterial coat and may lead to thrombosis Usually only the smaller branches are involved Since it is a part of a pancarditis the diagnosis usually is not possible during life

CONGENITAL ANOMALY OF CORONARY ARTERY—See Congenital Anomaly of Heart or Great Vessels

17 EMBOLISM OF CORONARY ARTERY—A rare condition the symptomatology of which is due to myocardial infarction It occurs most often in the presence of acute or subacute bacterial endocarditis The diagnosis may be considered when death or signs and symptoms of myocardial infarction occur suddenly in an individual with bacterial endocarditis

18 OTHER DISEASES OF CORONARY ARTERY (specify)—Under this heading may be listed such diagnoses as are not mentioned elsewhere e g thrombo-angiitis obliterans aneurysm of coronary artery and the arteriolar lesions encountered in sickle cell anemia and lupus erythematosus

19 PERIARTERITIS NODOSA OF CORONARY ARTERIES—(Essential polyangiitis)—A rare condition not often diagnosed clinically It is part of a widespread involvement of the smaller or medium sized arteries and is of unknown etiology The diagnosis may be suspected when the anginal syndrome or signs of myocardial ischemia or infarction appear in the course of the systemic disease The electrocardiogram may present changes consistent with the myocardial involvement

20 STENOSIS OF CORONARY ORIFICE This condition is most often the result of syphilitic aortitis but may occasionally be found with arteriosclerosis of the supravalvular aorta The signs and symptoms are the same as described under Arteriosclerosis of Coronary Arteries When a lumen of an orifice has occurred sudden death may occur or the signs and symptoms of myocardial ischemia or infarction may follow

21 THROMBOSIS OF CORONARY ARTERY—This may or may not give rise to signs and symptoms depending upon the size and location of the vessel occluded and the state of the collateral circulation When it does give rise to symptoms they are those of myocardial ischemia or of myocardial infarction

forms on an arteriosclerotic or inflammatory lesion Some pulmonary thrombi propagate from an embolus

The onset of symptoms is not usually as abrupt as in embolism and *circulatory failure is less pronounced and less frequent* The signs and symptoms are otherwise similar to those described for Embolism of Pulmonary Arteries

Roentgenographic examination may reveal infarcts of the lungs which are often multiple and irregularly distributed They may be completely obscured by the accompanying pulmonary congestion

## DISEASES OF CORONARY ARTERIES

15 Arteriosclerosis of Coronary Arteries —This is usually part of a general arteriosclerosis but the coronary arteries may be the only ones affected Diagnostic signs and symptoms occasionally may be absent even when arteriosclerosis is present to a marked degree Arteriosclerosis of the coronary arteries results in narrowing of the lumen and consequent reduction of blood flow Coronary insufficiency (coronary failure) is the usual result of this and indicates that coronary blood flow has failed to keep pace with the myocardial needs so that myocardial ischemia is present

Symptoms may arise because of coronary insufficiency and this may come as a result of slowly progressive narrowing of the lumen or may be precipitated by an extracardiac event such as severe hemorrhage or exertion It may be manifested by

Anginal syndrome
Cardiac arrhythmia
Cardiac insufficiency
Electrocardiographic abnormalities as later specified under specific diagnoses of myocardial disease

When sufficiently prolonged or severe coronary insufficiency may give *rise to sudden death or to myocardial infarction*

Coronary occlusion may occur during the course of coronary arteriosclerosis The signs and symptoms vary according to the abruptness of the event the size and location of the affected myocardial area and the effectiveness of the collateral circulation With gradual onset no symptoms may appear or with abrupt onset they may be those of myocardial infarction

*Enlargement of the heart if present is due to a complicating factor* such as hypertension myocardial infarction or cardiac insufficiency

16 Arteritis of Coronary Arteries —This unusual lesion is found occasionally in rheumatic fever in thromboangiitis obliterans and

*Signs*

Pallor due to anemia
Embolic phenomena (petechiae Osler's nodes hematuria)
Clubbing of fingers
Evidence of valvular deformity or congenital anomaly
Enlarged spleen
Fever
Leucocytosis
Positive blood culture (The diagnosis occasionally may be made on clinical grounds in the absence of a positive blood culture)

26 Mural Endocarditis — This is a common finding in subacute bacterial endocarditis and in atypical verrucous endocarditis and occasionally in rheumatic endocarditis It also may complicate a ventricular septal defect It is chiefly of pathological interest for it cannot be recognized clinically

27 Mural Thrombosis —Thrombi may form on the endocardium of any of the chambers of the heart but are most frequently found in the atria and their auricles Among the causative factors are inflammation necrosis sclerosis and retardation of the blood flow as occurs with cardiac insufficiency

Mural thrombi are often friable and therefore commonly cause emboli either in the lungs or in the systemic circulation Rarely a *pedunculated* thrombus may become detached from the endocardium of the right or left atrium and occlude intermittently the tricuspid or mitral orifices (*ball valve thrombus*)

Atrial mural thrombosis is commonly encountered in rheumatic heart disease particularly in association with active rheumatic infection and also with atrial fibrillation and mitral stenosis Ventricular mural thrombi are most often encountered in myocardial infarction when it has involved the endocardium

In the presence of atrial fibrillation myocardial infarct cardiac insufficiency mitral stenosis or recurrent rheumatic activity with old valvulitis the diagnosis can be made from the following

*Signs*

Embolic manifestations in the pulmonary or systemic circulation or both
Attacks of syncope in a patient with mitral stenosis should suggest ball valve thrombus

22 TRAUMA OF CORONARY ARTERY (specify character of lesion) This is usually the result of a stab or gun shot wound It may also occur as an accident in paracentesis of the pericardial sac and in crush injuries of the chest It results in hemopericardium

## DISEASES OF ENDOCARDIUM AND VALVES

23 ENDOCARDITIS ACUTE BACTERIAL (specify organism)—This includes all forms of bacterial infection of the endocardium and valves Signs of sepsis usually *outweigh those of cardiac involvement* unless acute perforation of a valve occurs The diagnosis can be made when an acute infection is followed by

*Signs*

- Signs of sepsis
- Embolic phenomena
- The appearance of a cardiac murmur while the patient is under observation especially if it be a diastolic murmur or a change in the character or intensity of a pre existent murmur
- Positive blood culture

24 ENDOCARDITIS INDETERMINATE—Several different types of endocardial lesions are included under this heading Although the etiology of these is unknown they are probably not due to direct bacterial infection for the lesions do not contain bacteria They are mainly of pathologic interest One type however may occasionally be recognized clinically namely atypical verrucous endocarditis because of its frequent association with lupus erythematosus disseminatus

25 ENDOCARDITIS SUBACUTE BACTERIAL (Endocarditis Lenta) (specify organism)—This form is usually due to the streptococcus viridans more rarely the gonococcus the hemophilus influenzae or one of a variety of other bacteria It is usually engrafted upon old rheumatic valvular *deformities or congenital anomalies* more rarely upon sclerotic valves due to syphilis or arteriosclerosis Occasionally active rheumatic infection may co exist

Cardiac insufficiency rarely occurs early in the course of the disease but is a common terminal event It often causes death in patients who have *been cured of their* active infection Diffuse glomerulonephritis may be present and rarely may be the cause of death

Signs

- Apex beat heaving displaced to the left and downward
- Collapsing pulse
- Large pulse pressure with lowered diastolic and elevated systolic pressure
- A diastolic murmur high pitched and blowing in character occasionally musical It may if faint be best heard with the ear directly applied to the chest It is heard better with the diaphragm chest piece of the stethoscope than with the bell type It is usually loudest in the third or fourth interspace at the left border of the sternum or in the second or third interspace at the right border of the sternum It may at times be heard best in the apical area It is best heard with a relatively slow heart rate with the breath held in expiration and with the patient standing and bent forward
- A systolic murmur may be present at the aortic area in the absence of aortic stenosis when aortic insufficiency is marked In cases due to syphilis it is sometimes harsh and accompanied by a systolic thrill
- Roentgenological examination almost always shows enlargement of the left ventricle There is usually dilatation of the ascending aorta and increased amplitude of pulsations
- Electrocardiographic evidence of left ventricular hypertrophy is usual in advanced cases (Fig 40b Electrocardiographic Section)

b *Aortic stenosis*—This valvular lesion is usually associated with aortic insufficiency but may occur as an isolated lesion especially in the older age group It is often associated with the anginal syndrome Syncope or sudden death may occur Slight degrees of anatomical stenosis are *not* diagnosable but the diagnosis should be made on the basis of a characteristic murmur in combination with an anacrotic pulse

Signs

- Apex beat heaving displaced to the left and downward
- Systolic thrill best felt in the right first or second interspace close to the sternum It is elicited more *readily with the breath* held in expiration and the patient standing bent forward The presence of a thrill is not necessary for the diagnosis of aortic stenosis A systolic thrill may be felt over the carotid artery
- Loud harsh systolic murmur best heard in the right first or second interspace close to the sternum transmitted to the neck and widely over the precordium
- Anacrotic pulse with slow rise and broad summit

28 NEOPLASM OF ENDOCARDIUM—Myxomata arise from the endocardium, most often in the region of the fossa ovalis of the left atrium but sometimes from the valves They vary in size from a few millimeters to as much as six centimeters in diameter They are usually attached to a short stalk These tumors are of interest clinically because their size situation and pedunculated nature occasionally allow them to block intermittently an orifice of the heart usually the mitral causing syncope

Fibromas are endocardial tumors which arise from the subendothelium of the valves They do not produce clinical symptoms or functional disturbances

29 OTHER STRUCTURAL DISEASES OF ENDOCARDIUM OR VALVES (specify location if possible)—This includes conditions not mentioned in any of the other categories for example subendocardial fibrosis tuberculous endocarditis endocardial blood cysts and endocardial pockets

30 RUPTURE OF VALVE OR OF CHORDAE TENDINEAE (specify valve)—*The commonest cause of spontaneous rupture of a valve leaflet or of chordae tendineae is bacterial endocarditis* especially the acute form

*Rupture of a diseased valve* or of chordae tendineae may result from a sudden severe effort Sudden dyspnea *following unusual effort may arouse the suspicion that a valve has ruptured The occurrence of a* murmur indicating valvular deformity following such an episode or a change *in the* character of a murmur previously present confirm the diagnosis

31 TRAUMATIC INJURY OF ENDOCARDIUM OR VALVES (*specify lesion*)—Sudden compression of the thorax may cause rupture or tearing of a valve or of the endocardium of the atria A penetrating wound of the heart may involve the endocardium or the valve The diagnosis usually is made only at necropsy

32 VALVULAR *DEFORMITY designates the result of acute or chronic* disease affecting the heart valves Rheumatic valvulitis is the *common*est Less frequent are deformities due to syphilis arteriosclerosis and bacterial endocarditis Certain signs may aid in the diagnosis

a *Aortic insufficiency*—When the lesion is marked it may be associated with the anginal syndrome The diagnosis of aortic insufficiency should be made only in the presence of a characteristic murmur Cardiac enlargement rarely is absent

*Signs*

Apical impulse is brief sharp and not heaving

Diastolic thrill in the apical area

Heaving systolic pulsation to the left of the sternum between the third and fifth interspaces

The characteristic murmur of mitral stenosis is a low pitched presystolic or early diastolic rumble located at the cardiac apex The presystolic murmur often gives the impression of a crescendo character when the first sound is loud In the presence of atrial fibrillation with bradycardia the murmur is often early diastolic and has a diminuendo character Often it is heard throughout *diastole* At times the characteristic murmur can be elicited only after exercise or by having the patient lie on his left side

Loud snapping first sound at the apex The presence of such a sound together with the murmur of mitral insufficiency should arouse suspicion of mitral stenosis

The second sound at the pulmonic area is accentuated often reduplicated

A loud reduplication of the second sound *is often heard* over the body of the heart

Roentgenological examination reveals enlargement of the left atrium and right ventricle and often dilatation of the pulmonary artery Calcification of the mitral valve is sometimes observed

The electrocardiogram may show right axis deviation of QRS and notched broad large P waves In advanced cases *signs of right* ventricular hypertrophy or complete or incomplete right bundle branch block are often observed in precordial leads

e *Tricuspid insufficiency* —
f *Tricuspid stenosis* — } Structural deformities of the tricuspid valve though rarely recognized clinically are not uncommonly seen in rheumatic hearts at necropsy Deformities severe enough to cause insufficiency or stenosis or both are not as frequent Invariably they are associated with other valvular deformities especially *mitral stenosis* Occasionally bacterial endocarditis involving the tricuspid valve may produce insufficiency or *stenosis* Congenital *tricuspid* stenosis or atresia is discussed elsewhere

The clinical diagnosis of tricuspid valve deformity although difficult can be made correctly *in* an appreciable number of instances It should not be made in the presence of congestive heart failure since then tricuspid incompetency *is frequently* present *It* should be suspected in a rheumatic patient with mitral stenosis who has persistently enlarged liver engorged neck veins and recurrent ascites but who is free

- Low systolic blood pressure and small pulse pressure unless aortic insufficiency or essential hypertension is present
- Faint or absent aortic second sound
- Roentgenological examination reveals enlargement of the left ventricle and often calcification of the aortic valves
- Electrocardiographic evidence of left ventricular hypertrophy sometimes of left bundle branch block

c *Mitral insufficiency*—Deformity of the mitral valve gives rise to mitral insufficiency Rheumatic valvulitis is a common cause Sclerosis of valve flaps or of chordae tendineae may be due to atheromatous changes and cause mitral insufficiency It is important to bear in mind that incompetence of the valve ring due to myocardial disease may give rise to similar signs (mitral incompetency) and that apical systolic murmurs often exist without organic heart disease being present

The diagnosis should be made on the basis of a characteristic murmur combined with the history or evidence of an appropriate etiologic agent

*Signs*

- The apex beat may be forceful and displaced to the left and downward
- The characteristic sign is a systolic murmur best heard in the apical area often obscuring the first sound and usually transmitted to the left axilla The characteristic features of this murmur are its long duration and moderate or loud intensity Its quality may be blowing harsh or musical and of relatively high pitch
- Enlargement of the heart may be considerable It is often slight and recognizable only by roentgenological methods
- Loud pulmonic second sound
- Roentgenological examination reveals enlargement of the left ventricle and atrium in more advanced stages enlargement of the right chambers as well

d *Mitral stenosis*—The diagnosis should be based primarily on the presence of a characteristic murmur and on evidence of enlargement of the left atrium Mitral insufficiency usually is also present

In a few cases of aortic insufficiency a diastolic murmur is heard at the apex simulating the murmur of mitral stenosis (Austin Flint murmur) If aortic insufficiency is of rheumatic origin a rumbling diastolic murmur at the apex is likely to be due to mitral stenosis If the aortic insufficiency is syphilitic it is fair to assume that the mitral valve is intact

*Signs*

Apical impulse is brief sharp and not heaving

Diastolic thrill in the apical area

Heaving systolic pulsation to the left of the sternum between the third and fifth interspaces

The characteristic murmur of mitral stenosis is a low pitched pre systolic or early diastolic rumble located at the cardiac apex The presystolic murmur often gives the impression of a crescendo character when the first sound is loud In the presence of atrial fibrillation with bradycardia the murmur is often early diastolic and has a diminuendo character Often it is heard throughout diastole At times the characteristic murmur can be elicited only after exercise or by having the patient lie on his left side

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f *Tricuspid stenosis* — } Structural deformities of the tricuspid valve though rarely recognized clinically are not uncommonly seen in rheumatic hearts at necropsy Deformities severe enough to cause insufficiency or stenosis or both are not as frequent Invariably they are associated with other valvular deformities especially mitral stenosis Occasionally bacterial endocarditis involving the tricuspid valve may produce insufficiency or stenosis Congenital tricuspid stenosis or atresia is discussed elsewhere

The clinical diagnosis of tricuspid valve deformity although difficult can be made correctly in an appreciable number of instances It should not be made in the presence of congestive heart failure since then tricuspid incompetency is frequently present It should be suspected in a rheumatic patient with mitral stenosis who has persistently enlarged liver engorged neck veins and recurrent ascites but who is free

of orthopnea Murmurs rarely contribute to the diagnosis of this valvular deformity since it is difficult or impossible to distinguish them from the murmurs of mitral stenosis and insufficiency which are almost invariably present in these patients

*Signs of tricuspid valve disease*

Cyanosis often combined with icterus
Distended neck veins which may show unusually marked pulsations
Orthopnea is rare
Atrial fibrillation is frequent
Enlarged liver which may show unusually marked pulsations
Recurrent ascites
Roentgenologic examination will show enlargement of the right atrium and unexpectedly clear pulmonary fields
Right axis deviation of QRS is common

*Signs suggesting tricuspid insufficiency*

Systolic pulsation of the cervical veins and often of the veins of the extremities
Systolic expansile pulsation of the liver

*Signs suggesting tricuspid stenosis*

Forceful presystolic pulsation of the neck veins
Expansile pulsation of the liver preceding the radial pulse
Ascites with minimal edema of the legs

33 VALVULAR SCLEROSIS (specify valve affected) —By this term is meant involvement and deformity of a valve by arteriosclerosis or other degenerative processes In the more advanced stage calcification is present The aortic and mitral valves are most commonly affected Occasionally the rings of these valves are also involved producing rigid collar like lesions

The mitral leaflets especially the anterior one are often calcified but since only portions of the leaflets are involved function usually is not disturbed The condition may be suspected clinically in the presence of a loud harsh or musical systolic murmur in the apical area with or without a *palpable thrill*

In elderly subjects the aortic valve may be sclerotic calcified and deformed the mitral valve frequently remaining normal This may result in stenosis of the orifice with or without insufficiency of the valve The physical signs of aortic stenosis aortic insufficiency or both may be present It is often impossible to decide whether the calcifica

tion is due to a degenerative process (Monckeberg type of arteriosclerosis) or is the result of an old rheumatic valvulitis. Usually in the former there are no signs referable to the mitral valve and a history of rheumatic infection is lacking. The degree of deformity of the aortic valve determines the presence or absence of enlargement of the heart.

The presence of calcium in these valvular lesions may be demonstrated by appropriate roentgenologic techniques.

34 Valvulitis, Active (specify deformity if any).—Active valvulitis is most commonly observed as a part of rheumatic endocarditis, though bacterial endocarditis and syphilis also may involve the valvular structure.*

The process may be either acute or chronic, the differentiation being chiefly on the basis of the duration of the inflammation. Chronic active valvulitis implies continued activity of the infection within the valve over a long period of time, the exact duration not being prescribed.

Acute rheumatic valvulitis is one of the commonest lesions encountered in the heart in rheumatic fever. It is then usually *part of a pancarditis in which the myocardium, endocardium, valves and pericardium are involved.* *

Both the endocardium and the supporting tissues of the valves are affected. The valve most commonly involved is the mitral, less often the aortic, and occasionally the tricuspid and pulmonary valves.

In the acute stage of rheumatic valvulitis the changes are not of sufficient degree to cause deformity of the valve orifice or dysfunction of the valve. The associated myocarditis, together with the acute inflammatory changes in the ring portion of the valve, however, may produce dilatation of the valve ring with resultant valvular incompetency and enlargement of the heart. Thus at best the diagnosis of acute rheumatic valvulitis is an inferential diagnosis made from certain physical signs which may have various causes and which may disappear with the acute stage. If they persist we may conclude that valvular deformity has developed.

*Acute active mitral valvulitis* may be suspected in the presence of a systolic murmur, usually blowing in character and localized at the cardiac apex. Occasionally, especially in children, a transient low pitched mid-diastolic murmur of unknown cause may be heard in the apical area.

* The use of "active" and "inactive" refers to the etiological agent; "acute," "chronic," "healing" and "healed" apply to the anatomical state.

* Owing to the large number of tissues affected and the similarity of the signs produced by disease of each one, a diagnosis of pancarditis must always be considered.

of orthopnea Murmurs rarely contribute to the diagnosis of this valvular deformity since it is difficult or impossible to distinguish them from the murmurs of mitral stenosis and insufficiency which are almost invariably present in these patients

*Signs of tricuspid valve disease*

Cyanosis often combined with icterus
Distended neck veins which may show unusually marked pulsations
Orthopnea is rare
Atrial fibrillation is frequent
Enlarged liver which may show unusually marked pulsations
*Recurrent ascites*
Roentgenologic examination will show enlargement of the right atrium and unexpectedly clear pulmonary fields
Right axis deviation of QRS is common

*Signs suggesting tricuspid insufficiency*

Systolic pulsation of the cervical veins and often of the veins of the extremities
Systolic expansile pulsation of the liver

*Signs suggesting tricuspid stenosis*

Forceful presystolic pulsation of the neck veins
Expansile pulsation of the liver preceding the radial pulse
Ascites with minimal edema of the legs

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In elderly subjects the aortic valve may be sclerotic calcified and deformed the mitral valve frequently remaining normal This may result in stenosis of the orifice with or without insufficiency of the valve The physical signs of aortic stenosis aortic insufficiency or both may be present It is often impossible to decide whether the calcifica

a previous infarction are present Persistent abnormal S T elevation in the standard and chest leads is not uncommon

37 ATROPHY OF HEART —This condition is associated with chronic wasting diseases and is of no clinical importance Usually the diagnosis is made only at necropsy

38 CONGENITAL ANOMALY OF HEART OR GREAT VESSELS (specify lesion if possible)—Certain congenital cardiovascular anomalies are easily diagnosed by a careful analysis of the history physical signs roentgenologic and electrocardiographic studies Other cases with malformations which are usually multiple may need further study by angiocardiography cardiac catheterization etc. Not all of the congenital anomalies are considered in this text Those not specifically mentioned may be filed under the above title as may also those that are undiagnosed

## A NON CYANOTIC GROUP

A 1 ANEURYSM OF SINUS OF VALSALVA CONGENITAL —Caused by a weakness in the aortic septum usually at the base of the right sinus of Valsalva It usually is recognized only after the aneurysm has ruptured into the right atrium or ventricle with sudden onset of heart pain and dyspnea and progressive right heart failure without cyanosis

*Signs*

Sudden appearance of continuous loud murmur with diastolic accentuation and thrill in the third and fourth left intercostal spaces

Roentgenographic demonstration of progressive increase in size of the right atrium or ventricle Occasionally the aneurysm may be demonstrated by angiocardiography before it has ruptured

A 2 AORTIC RING DOUBLE AORTIC ARCH —Persistence of both the right and the left fourth branchial arches These aortic arches join posteriorly to form the descending aorta thereby encircling the trachea and usually also the esophagus Symptoms may be absent if the vascular ring is large enough not to encroach upon the enclosed trachea and esophagus but if it is small respiratory distress and dysphagia are severe

*Signs*

(If vascular ring is small) Increased respirations with suprasternal and intercostal retractions

In the presence of *acute active aortic valvulitis* a high pitched diastolic murmur like that of aortic insufficiency may occur This murmur also is often of transitory nature and is more likely to occur in children

In the presence of *chronic active valvulitis* progressive valvular deformity of permanent nature usually occurs The murmurs described above become permanent and assume the characteristics mentioned under Valvular Deformity

In most instances of active rheumatic valvulitis clinical manifestations of disease of other cardiac tissues will be present especially those due to inflammation of the myocardium (See Myocarditis Acute) There also will be evidence of activity of the etiologic factor

*Signs*

The appearance of a murmur or murmurs systolic or diastolic at the apex or aortic area during the course of rheumatic fever

Slight or moderate enlargement of the heart

Evidence of active myocarditis or pericarditis

35 VALVULITIS INACTIVE (specify deformity if any) —Recovery from acute valvulitis may *occasionally* be *complete* and without deformity usually there is localized or extensive scarring

The diagnosis of healed valvulitis is based on finding the signs of valvular deformity without evidence of activity of the etiologic factor

## DISEASES OF MYOCARDIUM
### (Including Conduction System and Heart as a Whole)

36 ANEURYSM OF HEART (specify location) —This lesion usually is the result of myocardial infarction The apical portion of the left ventricle is most commonly affected (See Pathological Section) The diagnosis is based upon physical signs and roentgenographic findings aided by a history of previous infarction There are no characteristic symptoms

*Signs*

Abnormal precordial systolic pulsation mesial to and above the apex impulse may be observed in marked cases

Faint heart sounds at the apex

Fluoroscopic finding of a localized bulge usually with systolic expansion Sometimes also the diastolic movement will be opposite to that of the remainder of the ventricular outline

Electrocardiogram not characteristic except when features suggesting

the branches of the aorta given off above and those given off below the constriction. The lesion is found more frequently in males. Development is normal. The complexion is usually florid.

*Signs*

Heart is usually enlarged
Systolic murmurs especially at the base of the heart, in the interscapular regions and in both axillae
Thrills may be felt in the same areas
Pulsations of the distended vessels of the collateral circulation may be felt
Absent or diminished femoral pulsations
Blood pressure higher in upper extremities than in lower
Roentgenologic examination may show: Slight or moderate left ventricular enlargement, dilated ascending aorta with small aortic knob, poorly visualized descending aorta, notching along lower margins of ribs (not commonly present before puberty)
(Fig. 33 A, B, C of the Roentgenological Section)
Angiocardiography may locate the stenotic segment and indicate its extent and length. It may indicate whether post-stenotic aortic dilatation is present or whether the left subclavian artery is involved, and demonstrate the collateral arterial anastomoses
(Fig. 33 D of the Roentgenological Section)

A 6. DEXTROCARDIA.—Due to a developmental anomaly in which the primitive cardiac loop is reversed. The heart lies in the right chest with the apex pointing to the right, a mirror image of the normal position of the heart. Dextrocardia is usually accompanied by situs inversus or it may be accompanied by grave cardiac lesions.

*Signs* (in uncomplicated dextrocardia)
Asymptomatic
Apex thrust seen and felt in fourth or fifth right intercostal spaces
Heart sounds heard better over right chest
Electrocardiogram shows an inversion of all deflections in Lead I with Lead II resembling Lead III and vice versa
Roentgenographic examination shows a reversed picture of the heart with the apex to the right

A 7. DILATATION OF PULMONARY ARTERY (PRIMARY).—This may be differentiated from the post-stenotic dilatation of the pulmonary artery by the fact that the pulmonic second sound is accentuated and that on

Brassy cough with stridor
Dysphagia
Roentgenologic examination shows a wide mediastinal shadow
Angiocardiography will reveal the vessels concerned in the constriction

A 3 AORTIC STENOSIS—A congenital thickening of the aortic valves is present The symptoms usually are minimal although occasionally syncope or sudden death may occur Subacute bacterial endocarditis is a common complication

*Signs*
As described for Aortic Stenosis (See under Valvular Deformity)

A 4 BICUSPID AORTIC VALVE—This lesion has no characteristic clinical signs It may be suspected in the presence of an infective endocarditis of the aortic valve when causes of acquired valvular disease can be excluded It should also be suspected when an aortic diastolic murmur is found in an otherwise normal young individual with negative rheumatic history or in a patient with coarctation of the aorta

A 5 COARCTATION OF AORTA—May be of the infantile or adult type
(1) *Infantile type*—There is a diffuse narrowing of the isthmus of the aorta between the origin of the left subclavian artery and the ductus arteriosus The narrowing also may involve the left subclavian and occasionally the left carotid artery The ductus arteriosus remains patent so that blood from the pulmonary artery passes into the aorta beyond the narrowing The infantile type is rare and difficult to diagnose and is usually fatal as an isolated lesion It is important to recognize it because surgery may be helpful

*Signs*
Slight cyanosis in lower extremities while ductus remains open
A weak pulse in lower extremities
Angiocardiography may show opacity within the ductus arteriosus and the descending aorta before the filling of the left ventricle and aortic arch

(2) *Adult type*—There is a localized constriction of the aorta usually beyond the origin of the left subclavian artery and at or near the insertion of the ductus arteriosus The ductus arteriosus closes normally so that there is a resulting compensatory collateral circulation between

Roentgenographic demonstration of normal heart size at birth followed in subsequent examinations by rapidly progressive left ventricular enlargement and diminished amplitude of left ventricular pulsations

A 11 PULMONARY VEINS ALL DRAIN INTO RIGHT ATRIUM —The veins empty either directly or through the coronary sinus to the right atrium or deviously through a persistent left superior vena cava and a left innominate vein into the right superior vena cava The left atrium and ventricle are small as they can only receive blood through an atrial septal defect Life is dependent upon an adequate interchange of blood from right to left through such a defect

*Signs*

Heart size normal at birth showing progressive rapid enlargement of the right side

Blood pressure usually low

Oxygen content of blood in the right atrium is identical with that in the arterial circulation

Heart failure eventually develops

A 12 PULMONARY VEINS ENTERING RIGHT ATRIUM OR SUPERIOR VENA CAVA —One or more veins may be involved the remainder draining into the left atrium While the right side of the heart receives more blood and the left side less than normal there is little obvious change in the circulation

*Signs*

Roentgenologic examination may show dilatation of the superior vena cava a prominent pulmonary artery and enlargement of the right atrium

The anomalous veins may be demonstrated by angiocardiography

Catheterized blood sample from right atrium will show higher oxygen content than expected

A 13 PULMONIC STENOSIS ISOLATED —May be valvular or infundibular in type In the valvular type there is fusion of the three semilunar cusps which form a diaphragm or cone shaped dome A small aperture in this leads to the post stenotic dilated pulmonary artery In the infundibular type the pulmonary conus is involved There is an arrest in the involution of the bulbus cordis so that the right ventricle is divided by a muscular ridge into a main chamber and an infundibular one leading to the pulmonary artery

cardiac catheterization the systolic pressure is not elevated in the right ventricle but is higher than in the pulmonary artery The diagnosis of this condition depends on the exclusion of *pulmonic or infundibular* stenosis by cardiac catheterization

*Signs*

Harsh systolic murmur best heard in the second left intercostal space
Systolic thrill best felt in this area
Pulmonary second sound is increased
*Electrocardiogram* shows the features commonly found with right ventricular hypertrophy
Circulation time normal
On cardiac catheterization the systolic pressure in the right ventricle is not elevated but is greater than in the pulmonary artery
On roentgenologic examination there is pulmonary artery dilatation without right ventricular enlargement
Angiocardiography shows the variations in the degree and extent of the dilatation of the pulmonary artery and its branches

*A 8* HYPERTROPHY OF HEART CONGENITAL —*Massive enlargement* of the heart present at birth not associated with other cardiac anomalies The condition is fatal in early months of life

*Signs*

Dyspnea
Faint heart sounds
*Roentgenologic evidence of marked general enlargement of heart*

A 9 HYPOPLASIA OF AORTA —A developmental arrest in the normal growth of the aorta rarely found as an isolated lesion More commonly it is secondary to an anomaly in which the blood flow to the aorta is decreased as atrial septal defect or aortic stenosis

A 10 LEFT CORONARY ARTERY ARISING FROM PULMONARY ARTERY — These patients may be asymptomatic for the first few months of life and then usually show progressive signs and symptoms of left ventricular failure Death usually occurs before the 6th month but rarely there may be a normal span of life

*Signs*

Electrocardiogram may *show inversion of T waves in Leads I and II* and in precordial leads over the left ventricle

the left pressure may be caused by the ligamentum arteriosum or by a left subclavian artery arising from the left aortic root Usually the vascular ring so formed is wide enough not to cause symptoms but if small it may encroach upon the enclosed trachea or esophagus or both causing signs of compression These cases show a tendency to pulmonary infections They often have a brassy cough with stridor and occasionally have dysphagia

*Signs*

Roentgenologic examination reveals an aortic knob on the right
There is displacement of the esophagus and less frequently of the trachea to the left and anteriorly (Fig 31 of the Roentgenological Section)
Angiocardiography helps to visualize the position of the aorta

A 15 SUBAORTIC STENOSIS —Due to an arrest in the involution of the bulbus cordis a shelf of fibrous tissue persists stretching below the aortic valves The lesion is found more frequently in males The symptoms are minimal Subacute bacterial endocarditis is a common complication

*Signs*

As described for Aortic Stenosis (See under Valvular Deformities) except that the aortic second sound is not diminished
Angiocardiography may show narrowing of the isthmian portion of the left ventricle

A 16 TRICUSPID VALVES IN ANOMALOUS POSITION —The posterior leaflet is displaced downward into the right ventricle (Ebstein's disease) Right atrial enlargement is present and tricuspid insufficiency appears terminally These patients are often asymptomatic but dyspnea and palpitation and even sudden death may occur Arrhythmias and right ventricular hypertrophy are frequent

## B POTENTIALLY CYANOTIC GROUP
(Cyanose tardive)

Anomalies of the atrial septum are the most common congenital heart lesions found in the adult

B 1 ATRIAL SEPTAL DEFECT —This lesion constitutes a failure in the development of the atrial septum It is usually partial involving the

Cyanosis is absent until congestive failure supervenes except when the foramen ovale is not completely sealed This allows a right left shunt and may cause slight cyanosis

*Signs*

There may be unusual prominence of the left anterior chest due to hypertrophy of the right ventricle
Harsh systolic murmur best heard in the 2nd left intercostal space
Systolic thrill best felt in the 2nd left intercostal space
Pulmonary second sound may be normal or decreased
Electrocardiogram shows the features commonly found with right ventricular hypertrophy sometimes right bundle branch block
Circulation time normal
On cardiac catheterization the systolic pressure in the right ventricle is elevated and that in the pulmonary artery is decreased
Roentgenologic examination reveals right ventricular and right atrial enlargement and except with infundibular stenosis dilatation of the pulmonary conus The branches of the pulmonary artery are small so that the lung fields are unusually clear
On angiocardiography the site of stenosis may be demonstrated

A 11 RIGHT AORTIC ARCH – A persistence of the fourth right branchial arch to form the aortic arch on the right with atrophy of the fourth left branch The three main vessels given off the arch are the mirror image of the normal

The aorta (1) may descend on the right of the spinal column or (2) it may cross abruptly behind the esophagus to descend in its usual position on the left

(1) *Right aortic arch with aorta descending on right* – There are no symptoms

*Signs*

Roentgenologic examination in the postero anterior position shows the aortic knob to the right of the sternum
This may be visualized as a denser shadow within that of the superior vena cava
In the left anterior oblique position the anterior aspect of the esophagus is indented by the aorta
With ingestion of barium the right margin of the esophagus is seen to be indented by the aortic arch

(2) *Right aortic arch with left descending aorta* – After forming the arch on the right the aorta curves abruptly behind the esophagus to

of the right atrium from the left may be demonstrated (Fig 28 of Roentgenological Section)

Oxygen content of the blood in the right atrium is greater than in the superior vena cava

B 2 COMBINED ATRIAL AND VENTRICULAR SEPTAL DEFECTS (persistent atrioventricularis communis) —This is due to an early arrest in the development of both the atrial and ventricular septa Because of the resulting defect the fused leaflets of the mitral and tricuspid valves form a single atrioventricular valve Rarely the defect may be so large that a single atrium and single ventricle are present (cor biloculare) The shunt is predominantly from left to right depending upon which chamber has the higher pressure

*Signs*

Oxygen saturation of arterial blood is low
Cyanosis is minimal
Systolic murmur and thrill at third and fourth left intercostal spaces to the left of the sternum
Oxygen content higher in right atrium than in superior vena cava
Roentgenologic examination shows progressive but moderate enlarge ment of the right atrium and ventricle

P 3 PATENT DUCTUS ARTERIOSUS —A persistence of the fetal shunt between the pulmonary artery and the aorta joining the latter beyond the *origin* of the left subclavian artery One of the most common con genital heart lesions Found more frequently *in females* Growth is retarded if the patency is large

*Signs*

The typical murmur is continuous harsh with systolic accentuation
It is heard best at the 2nd left intercostal space and is transmitted to the vessels of the neck
In rare cases the diastolic component may be absent
There is often a systolic thrill best felt in the 2nd left intercostal space
Diastolic blood pressure is low with *increased* pulse pressure
Roentgenologic examination usually shows prominence of the pul monary artery segment The hilar branches of the pulmonary artery may be dilated and pulsations of *increased* amplitude may be noted Left and right ventricular enlargement may be present but often is not. (Fig 32 in Roentgenological Section)

cephalic (upper) part of the septum or the basal part (persistent ostium primum) In the latter case the base of the opening is formed by the tricuspid and mitral valves If the atrial defect is complete it produces cor triloculare biventriculare Free communication exists between the atria with a left to right shunt occurring as long as the left atrial pressure is greater than the right Hypertrophy of the right side of the heart and dilatation of the pulmonary artery and its branches occur A reversal of blood flow with resultant venous-arterial shunt appears during physical strain and with right ventricular failure

The condition is more frequent in females The habitus is frail Lung infections and rheumatic carditis are common Atrial fibrillation is not rare in older patients Atrial septal defect is frequently associated with a stenosis of the mitral valve which may be congenital or an acquired rheumatic lesion This combination is called the Lutembacher syndrome

Subacute bacterial endocarditis is rare with this lesion

*Signs*

Transitory cyanosis appearing after physical strain

Development of cyanosis in a previously acyanotic individual

Pulmonic second sound is loud and ringing

Inconstant systolic murmur heard in the 2nd and 3rd intercostal spaces to the left of the sternum (4th and 5th spaces with persistent ostium primum)

A low pitched rumbling diastolic murmur of variable intensity may be heard at the apex (Lutembacher syndrome)

An inconstant systolic thrill may be felt near the sternum

Enlarged heart

Low blood pressure with small pulse pressure

Electrocardiographic signs of moderate right axis deviation in bipolar limb leads with large R or R' waves in precordial leads from the *right side of the heart* *Right bundle branch block is common* High sharply peaked P waves in Leads II and III

Roentgenologic examination may show marked enlargement of the right atrium and ventricle with prominent and markedly dilated pulmonary artery and branches The left ventricle is normal sized or small the aorta commonly is hypoplastic The left atrium may not be greatly enlarged even with coincident mitral stenosis Increased amplitude of hilar pulsations may be present (Fig 29 in Roentgenological Section)

On angiocardiography the opaque substance at times may pass immediately from the right to the left atrium Rarely reopacification

Electrocardiogram shows marked right ventricular hypertrophy pattern

Roentgenologic demonstration of marked right ventricular and atrial enlargement prominent pulmonary artery and dilated superior vena cava The small left ventricle may be identified in the left anterior oblique positions The small aortic arch may be demonstrated in oblique views

C 2 EISENMENGER COMPLEX —The aorta is dextroposed and over rides a high ventricular septal defect thus receiving blood from both ventricles The pulmonary artery is usually normal in size though it may be large The right ventricle is usually hypertrophied

*Signs*

Persistent cyanosis usually appears at puberty
Clubbing of fingers may appear later
Occasional hemoptysis
Heart size normal or slightly enlarged
Pulmonic second sound usually accentuated
Loud systolic murmur over pulmonic area
Systolic thrill over pulmonic area or somewhat lower
Circulation time is abnormally short
Roentgenologic examination usually reveals a heart of *normal size* though there may be right ventricular enlargement with rounding and elevation of the apex The pulmonary artery and its branches are usually *dilated* and there is increased pulsation of the hilar vessels The vascularity of the lungs usually is increased
Angiocardiography shows simultaneous filling of the pulmonary artery and aorta (Fig 30 of the Roentgenological Section)
The distinction between this condition and the tetralogy of Fallot with post stenotic pulmonary artery dilatation can be made only by the demonstration of significant pressure differences between the right ventricle and the pulmonary artery on cardiac catheterization in the latter condition

C 3 PERSISTENT TRUNCUS ARTERIOSUS —Due to failure in development of the aortic pulmonary septum a single common great vessel over rides a high ventricular septal defect and receives blood from both ventricles to supply the greater and also the lesser circulation The lungs are supplied by blood either (1) through pulmonary artery branches given off from the *truncus arteriosus* or (2) through the bronchial arteries

Oxygen content of the blood is higher in the pulmonary artery than in the right ventricle

B 4 PATENT FORAMEN OVALE (persistent ostium secundum) —Although functionally closed soon after birth by a valve which lies on the left side of the atrial septum the foramen ovale may remain open anatomically and not be considered an anomaly When however pressure in the right atrium becomes greater than that in the left the valve is forced open and a right to left shunt takes place Cyanosis may appear if sufficient blood passes into the left atrium Paradoxical emboli may occur under these conditions

B 5 VENTRICULAR SEPTAL DEFECT (maladie de Roger) —An opening in the ventricular septum usually small in size and near the basal part of the septum If the defect should be large in this situation the pulmonary artery may be dilated The condition is asymptomatic

*Signs*

- Loud harsh often high pitched systolic murmur best heard in the third and fourth left intercostal space near the sternum It often is not widely transmitted
- Electrocardiogram is usually normal but may show evidence of right ventricular hypertrophy
- The roentgenologic appearance is not characteristic It may be normal or the right ventricle may be enlarged and the pulmonary artery dilated
- Angiocardiography occasionally may demonstrate reopacification of the right ventricle

## C CYANOTIC GROUP

C 1 AORTIC ATRESIA OR STENOSIS —Left ventricle is underdeveloped With this lesion patency of the ductus arteriosus and atrial septal defect are essential for completion of the circulation

*Signs*

- Marked cyanosis
- Marked dyspnea
- Second sound at base not reduplicated
- Pulse in upper and lower extremities small and equal on the two sides
- Low blood pressure

anteroposterior position with no enlargement of the right ventricle in the left anterior oblique There is marked hilar congestion

C 5 TETRALOGY OF FALLOT —Pulmonic or infundibular stenosis combined with dextroposition of the aorta ventricular septal defect and right ventricular hypertrophy constitutes the tetralogy of Fallot a right aortic arch is often found with this condition

*Signs*

Poor development is the rule
Prominence of the left anterior chest
Children frequently assume the squatting position
Marked cyanosis uniform and persistent increased by exercise
Arterial thrombi may occur
Dyspnea on slight exertion
Clubbing of fingers and toes
Heart usually of normal size
Systolic murmur and thrill usually over pulmonic area in children In adults usually below this area
Polycythemia is present
The circulation time is shortened
Roentgenologic examination may reveal enlargement with rounding and elevation of the apex and a concave or inconspicuous pulmonary artery segment The hilar branches of the pulmonary artery are narrowed and the vascularity of the lung fields decreased There may be a normal or a dilated pulmonary artery and branches with normal pulsations and the right ventricle may not be demonstrably enlarged The presence of a right or left aortic arch can be established by the characteristic displacement of the esophagus
Angiocardiography reveals simultaneous opacification of the right chambers and the aorta The left ventricle and pulmonary artery may be visualized at the same instant Occasionally the site and degree of the stenosis may be demonstrated The size and location of vessels which might be utilized for anastomosis often are clearly seen (Fig 27 of the Roentgenological Section)
Electrocardiogram is consistent with marked right ventricular hypertrophy
Oxygen saturation of the arterial blood is low and is further decreased by effort

*Signs*

Cyanosis and dyspnea may be slight if pulmonary artery branches are given off the truncus arteriosus but are marked if the circulation to the lungs is through the bronchial arteries

Clubbing is present

The heart is enlarged

Second sound at the base is not accentuated

Systolic murmur and thrill loudest at the pulmonic area

Roentgenologic examination shows absence of the pulmonary artery contour and greatly enlarged right ventricle and right atrium

In the left anterior oblique position the upper horizontal margin of the right atrium makes an abrupt angle with the ascending aorta

C 1 SINGLE VENTRICLE WITH RUDIMENTARY OUTLET CHAMBER (*cor triloculare biatriatum*) —*An early arrest in the development of the heart* in which both the common ventricle and the bulbus cordis persist The latter is a rudimentary chamber from which one or both great vessels may arise or the great vessels may be transposed Various combinations of aortic and pulmonary artery dilatation or narrowing may occur *depending upon the origin of each from the common ventricle or the* rudimentary chamber

*Signs*

Heart size normal at birth showing gradual enlargement

Murmurs usually are not present

Oxygen saturation of arterial blood is decreased whether or not cyanosis is visible

Electrocardiogram may show predominant S wave in Leads I II and III

Roentgenologic examination may indicate moderate though progressive cardiac enlargement in successive roentgenograms

(1) *When pulmonary artery arises from rudimentary chamber*

*Signs*

Cyanosis is intense

Roentgenologic examination shows no prominence of the pulmonary conus and the pulmonary window is clear in the left anterior oblique position

(2) *When great vessels are transposed aorta arising from rudimentary chamber, pulmonary artery from common chamber*

*Signs*

Cyanosis absent

Roentgenologic examination shows dilated pulmonary conus in

39 DEGENERATION OF MYOCARDIUM (*specify variety if possible*) — Heart muscle is susceptible to toxins of bacterial chemical metabolic or vegetable origin Parenchymatous degeneration may occur in typhoid fever scarlet fever diphtheria and trichinosis In the latter two cases there may be an inflammatory reaction also Mineral poisons especially arsenic and phosphorus may cause severe degeneration of the heart muscle

Anemias both primary and secondary may cause fatty degeneration Degeneration of the heart muscle may result from an invasion of the myocardium by an inflammatory process A parenchymatous degeneration with disseminated patchy replacement fibrosis may be caused by protracted or recurrent myocardial ischemia as a result of coronary arteriosclerosis with blood flow insufficient for the functional requirements of the heart muscle

The diagnosis is usually made only at necropsy but should be suspected clinically in the presence of any of the aforementioned etiological agents

*Signs*

Sinus tachycardia
Abnormal rhythms
Faintness or sharp quality of the first heart sound
Systolic murmur at the cardiac apex
Enlargement of the heart
Evidence of cardiac insufficiency
Electrocardiogram may show abnormalities such as low voltage shallow or inverted T waves or defective conduction

40 ENLARGEMENT OF HEART (*chambers involved may be specified*) — This may be due to hypertrophy of the muscle or dilatation of the chambers It is usually due to a combination of both though one or the other may predominate Enlargement may be caused by conditions within the heart such as valvular deformities myocardial degeneration or prolonged abnormal rhythms or by constitutional disorders such as arterial hypertension severe anemia hyperthyroidism or beriberi A combination of causes is often present in a single individual

*Signs*

Apex beat forceful and covers a larger area than normal it may be shifted to the left or downward
Area of cardiac dullness increased in extent
Increased size of heart on roentgenologic examination Since slight

C 6 TRANSPOSITION OF GREAT VESSELS COMPLETE OR PARTIAL —The pulmonary artery lies behind the aorta and arises from the left ventricle while the aorta arises from the right ventricle In other cases only one vessel may have an abnormal origin The duration of life depends on associated lesions, but is usually short

*Signs*

Cyanosis progressively increasing during the first weeks of life
Dyspnea marked
Heart size normal at birth progressively increasing
Heart failure appears at an early age
Roentgenologic examination may reveal a narrow great vessel pedicle in the P A view Absence of the pulmonary arc in the P A view is significant There is progressive increase in size of the right ventricle and right atrium with increasing age
Angiocardiography may indicate the abnormal route of the blood flow In the lateral view the ascending aorta can be demonstrated to lie anterior to the pulmonary artery

C 7 TRICUSPID STENOSIS OR ATRESIA (pseudo triloculare) —A congenital anomaly due to malposition and irregular union of those parts of the cardiac septa dividing the mitral from the tricuspid ostium (Abbott) Defective development of the right ventricle and pulmonary artery are always present and unless blood can flow through an atrial septal defect and a patent ductus arteriosus the lesion is incompatible with life

*Signs*

Cyanosis persistent and of even distribution
Pulsation of the normal sized liver if the atrial defect is small
Systolic murmur occasionally present
Electrocardiogram usually shows left axis deviation of the QRS
Roentgenologic examination should demonstrate a *small right ventricle* an enlarged left ventricle absence of the pulmonary arc in the posteroanterior position a clear space beneath the aortic arch in the left anterior oblique position Absence of forward convexity of the right ventricle is characteristic
Angiocardiography demonstrates the flow from the right to the left atrium the right ventricle is poorly filled if at all The pulmonary artery usually fills from the aorta by way of a patent ductus arteriosus

On roentgenologic examination enlargement of the cardiac silhouette with general enlargement Enlargement of individual chambers may be recognized by an increased convexity of the peripheral border of the affected chamber

The *electrocardiogram* may show significant features especially in the chest leads In the presence of marked *right ventricular hypertrophy* leads from the right side of the precordium may present prominent R or R' waves and late intrinsicoid deflections associated occasionally with small Q waves and diphasic or inverted T waves In leads from the left side of the precordium small R waves may be associated with deep S waves In unipolar leads from the right arm large R deflections may be present

In *left ventricular hypertrophy* leads from the right side of the precordium show deep S waves and large upright T waves R deflections may be absent Leads from the left side present high R waves and late intrinsicoid deflections often associated with Q waves of conspicuous amplitude the S T segments are often depressed and the T waves may be diphasic or inverted

*Hypertrophy* of the *right atrium* may be associated with increased voltage of P in Leads II and III

*Hypertrophy* of the *left atrium* may be associated with a broad and sharply notched P wave of increased *duration* and perhaps also of increased voltage

*Marked ventricular hypertrophy* is often associated with high voltage in the bipolar leads and increased duration (*up to 0 12 sec*) of QRS These features are more often observed with left than with right ventricular hypertrophy Left ventricular hypertrophy is usually associated with left axis deviation of QRS and right ventricular hypertrophy usually with right axis deviation *These features* do not always accompany hypertrophy and further axis deviation of QRS is often observed in the absence of ventricular hypertrophy

Axis deviation may be absent even though ventricular hypertrophy is present Rarely it may show the reverse of the expected direction so that right ventricular hypertrophy may be associated with left axis deviation and left ventricular hypertrophy with right axis deviation

11 Fatty Infiltration of Heart —In obese individuals excessive deposits of adipose tissue may occur among muscle fibers or bundles especially of the right ventricle

Symptoms of cardiac insufficiency may be present but are usually due

or moderate enlargement of individual chambers may not be detected by roentgen examination limited to the frontal plane examination in both oblique positions is desirable observing the size and form of the individual chambers *

a *Dilatation*.—It is difficult to separate the signs of dilatation from those of hypertrophy since both conditions are commonly combined. Pure dilatation is infrequent and is usually an acute affair. It may be the result of myocardial degeneration or infection or anoxia and occurs as a response to increased pressure within a chamber. Dilatation may involve one chamber or multiple chambers. It is often accompanied by a systolic murmur due to relaxation of the mitral or tricuspid ring or both. The signs of dilatation will depend upon the chamber or chambers involved.

*Signs*

Apex beat usually absent or weak and displaced outward.

An increased area of *marked* cardiac dullness may be found, its situation depending upon the chamber or chambers affected.

Blowing systolic murmur at the apex.

Signs of cardiac insufficiency may be present.

Roentgenological examination shows a generally enlarged cardiac silhouette when the heart as a whole is dilated. Reduction in the size of the heart may be observed after treatment. When only one or two chambers are affected, prominent silhouettes of these chambers will be found.

b *Hypertrophy*.—The common causes of myocardial hypertrophy are arterial hypertension, valvular deformities and congenital malformations. In some hypertrophied hearts the etiology is unknown. Hypertrophy may be limited to one chamber or may involve two or more. Hypertrophy with little dilatation often cannot be recognized clinically. This is especially so in the concentric hypertrophy of the left ventricle due to arterial hypertension or aortic stenosis. Dilatation brings the hypertrophied heart closer to the anterior chest wall so as to produce diagnostic pulsations.

*Signs*

Forcible heaving apex beat due to hypertrophy of the left ventricle.

Systolic heaving of the precordial area adjacent to the left margin of the sternum due to hypertrophy of the right ventricle.

Loud booming first heart sound.

* If the prediction tables of Ungerleider and Clark (Table I) or of Hodges and Eyster (Table II) are used, enlargement is probable when the measurement of the heart exceeds the predicted measurement by as much as 10%.

coronary flow (coronary insufficiency) due to chronic narrowing of the lumen The precipitating factor in such cases lies either in a temporarily increased demand for coronary blood flow due to prolonged and unusual effort or in a diminished coronary flow due to shock or severe hemorrhage or prolonged tachycardia In patients with hypertension and marked cardiac hypertrophy the disproportion between muscle mass and vascular supply may afford a basis for infarction without occlusion Other rarer causes of infarction are Periarteritis nodosa rheumatic arteritis thrombo-angiitis obliterans embolism and syphilis

The location and extent of the infarct depend in part upon the position and caliber of the coronary arteries supplying the area and in part upon the adequacy of the collateral circulation to this area Infarcts may be localized in the anterior and apical portions of the left ventricle and the adjacent portion of the ventricular septum Infarction of the posterior and diaphragmatic portions of the left ventricle is also common and often includes the posterior portion of the septum The right ventricle is rarely affected by infarction except by extension from the adjacent septum

The extent of infarctions varies greatly It is commonly confined to the subendocardial region but may be transmural intramural or subepicardial A thin layer of focal necrosis in the subendocardium may result from prolonged and unusual effort hemorrhage or shock

*Symptoms*

Precordial pain or a sense of pressure is usually present It is of sudden onset with radiation to one or both arms or to the jaw It may start while at rest or during severe effort It is often described as pressing squeezing or crushing and is not relieved by nitroglycerin It is usually prolonged lasting a half hour or longer At times the pain is mild or even may not be present Occasionally pain may be epigastric and low sternal and this often is associated with nausea and vomiting Rarely the onset is with dyspnea and precordial oppression but without pain

*Signs*

Weakness sweating collapse
Pallor with or without cyanosis
Faint heart sounds
Gallop rhythm.
Pericardial friction rub
Cardiac arrhythmia

to coincident hypertension or coronary arteriosclerosis. Patients who are excessively obese may die suddenly without other signs of cardiac disease.

42. FIBROSIS OF MYOCARDIUM.—This term is employed to denote the condition in which the heart muscle shows diffuse fibrotic changes either due to arteriosclerotic or degenerative processes, or to acute or chronic inflammation. Clinically, it is usually found in individuals with arteriosclerosis of the coronary arteries or in those with arterial hypertension; less frequently in those with rheumatic heart disease or with myocarditis due to infections in which the myocardial inflammation has completely healed. Rarely it is found in those with syphilitic aortitis complicated by stenosis of the coronary ostia. It may be present without giving rise to signs or symptoms, though some degree of anginal syndrome or of cardiac insufficiency may be present. The diagnosis is chiefly by inference in the presence of an appropriate etiological factor.

*Signs*

- The first heart sound may be sharp or faint.
- Gallop rhythm.
- There may be a systolic murmur at the apex.
- Cardiac arrhythmia may be present, especially heart block or atrial fibrillation or ventricular premature beats.
- Enlargement of the heart may be present.
- The electrocardiogram may be normal or equivocal, or may show abnormalities of rhythm, of the QRS group, or the T wave, or of atrioventricular or intraventricular conduction. (See Electrocardiographic Section, Criteria for Interpretation.)

43. INFARCT OF MYOCARDIUM.—a. *Atrial infarction.*—Modifications of the P wave and the $T_1$ wave are not unusual in atrial infarction. The changes in the P wave are such as occur with disturbances of atrial rhythm. Often the apex of P is sharply pointed and may be of increased amplitude. The $T_1$ wave may be altered in a manner similar to that of the ventricular S-T segment in ventricular infarction.

b. *Ventricular infarction.*—This lesion is the result of a marked deficiency in coronary flow seriously affecting the nutrition of a localized portion of the ventricular myocardium. It may be due to thrombotic or atheromatous occlusion of a coronary artery. It may also occur without thrombosis or occlusion in an area of markedly diminished

Although localization of the infarct in the heart can be made in a gross way from the electrocardiogram exact anatomical definition may be quite inaccurate

44 MYOCARDITIS ACTIVE —Active myocarditis may be acute or chronic the differentiation being made arbitrarily on the basis of duration Chronic active myocarditis implies continued activity or frequent recurrence of the etiologic factor over a period of time Evidences of inflammation in the heart muscle are found most frequently in rheumatic fever occasionally in infectious diseases including subacute bacterial endocarditis in various septic states due to pyogenic bacteria and in other infections including those of viral origin In some forms of myocarditis the etiology is unknown Rheumatic fever is the most common cause of chronic active myocarditis The diagnosis is made largely by inference in the presence of an appropriate etiology

*Signs*

- Sinus tachycardia
- Abnormal rhythms
- Enlargement of the heart
- Systolic murmur at apex due to mitral incompetency
- Faintness or sharp quality of the first heart sound
- Evidence of cardiac insufficiency
- Electrocardiographic changes such as defective atrioventricular or intraventricular conduction or S T and T wave changes
- Fever
- Leucocytosis
- Increased sedimentation rate

45 NEOPLASM OF HEART (specify type) —Primary neoplasm is exceedingly uncommon and rarely diagnosed *during life* Metastatic neoplasm may be suspected in the course of malignant disease elsewhere in the body if cardiac symptoms or signs develop particularly atrial fibrillation or hemorrhagic pericardial effusion Occasionally primary tumors (myxoma) or secondary *neoplasms* arising from the wall of the atrium may grow in a polyp like *fashion* and obstruct the mitral or tricuspid ostium These may cause the signs of mitral or tricuspid stenosis There may be marked acute respiratory distress when in certain body postures obstruction of the ostium becomes nearly complete Electrocardiographic changes especially inversion of the T waves and persistent displacement of the S T junction and *segment* are occasionally observed in the presence of tumors in the ventricular myocardium

Basal rales or pulmonary edema or other manifestations of cardiac failure

Falling blood pressure sometimes after an initial period of elevation

Fever

Leucocytosis

Elevated sedimentation rate

Electrocardiographic records may reveal disturbances of rhythm and conduction The effects of local necrosis and ischemia may be apparent in the QRS deflections S T segment and T wave The particular features appearing will be influenced by the size location (spatial and mural) and age of the lesion and the degree of injury

The QRS group may have a low voltage in the *limb leads* Often with an infarct in the postero diaphragmatic area a large or relatively large $Q_3$ will appear less often a large $Q_1$ with anterior infarction Lead II may also show a large Q wave in either case In the *precordial extremity and esophageal leads* the QRS group may show changes of three principal types (1) An entirely negative deflection QS which may be notched or slurred to a variable degree (*this may be called the central type QRS* for it is obtained when the electrode is placed directly over the center of an experimental infarct (2) an abnormally deep or broad initial deflection Q followed by an R wave usually smaller than normal and occasionally also by S (this may be called the *marginal type QRS* for it is obtained when the exploring electrode is placed over the margin of an experimental infarct) (3) There may be no characteristic changes in QRS or simply a reduction in the size of the R wave

The S T junction and segment may be elevated or depressed This displacement is usually temporary Reciprocal deviation of the S T *segment is usually encountered in Leads I and III Lead I showing* elevation with anterior and Lead III with posterior infarction

*Subendocardial necrosis in the left ventricle may cause depression* of the S T segment of Leads I and II and in the left precordial leads and elevation in Lead $V_R$ and esophageal leads

Deviation of the S T segment usually gives place after a variable time to a T wave directed opposite to the original S T displacement

An abnormal form of the T wave may be the only evidence of recent myocardial necrosis These abnormalities may be transient or permanent If the former they remain for longer than do the S T segment abnormalities Because alterations in the S T segment and T wave are progressive several electrocardiograms may be necessary to demonstrate them

cardial infarction Hemopericardium is a common sequel of severe injuries of the heart Rupture of the heart may occur but usually is diagnosed only at necropsy

51 UNDIAGNOSED STRUCTURAL DISEASE OF HEART (specify location if possible) —This diagnosis should rarely be necessary

## DISEASES OF PERICARDIUM

52 CALCIFICATION OF PERICARDIUM —Occasionally *calcium is de*posited locally or diffusely in pericardial adhesions Calcification of the pericardium is often present without apparent embarrassment of the heart, or may accompany chronic constrictive pericarditis The diagnosis is made only by roentgenologic examination *especially in the* oblique or lateral views in which the calcium density is best observed (Figs 37 38 39 of the Roentgenological Section)

53 CONGENITAL ANOMALY OF PERICARDIUM (specify lesion) —This is a rare condition and usually symptomless

54 HEMOPERICARDIUM —Refers to the presence of blood in the *peri*cardial sac and does not include serosanguineous effusions It *is usually the result* of injury or rupture of the heart or aorta Clinically it is marked by a rapid progression of the diagnostic phenomena of cardiac tamponade

*Signs*

- Faint or absent heart sounds
- Increase in area of cardiac *dullness*
- Dullness over sternum replacing normal resonance
- *Increased* venous pressure
- Electrocardiogram shows abnormality *of* S T segment and of T wave
- Roentgenological *examinations reveal the* features mentioned under Serofibrinous Pericarditis

55 HYDROPERICARDIUM —This occurs as part of the anasarca of advanced cardiac insufficiency *or renal failure* or due to neoplastic infiltration The fluid is a transudate *The physical* signs are as given for serofibrinous pericarditis except that no friction rub is audible

56 NEOPLASM OF PERICARDIUM —Primary neoplasm of the pericardium is an extremely rare condition usually diagnosed only at

46 No Structural Disease of Heart

47 Other Structural Diseases of Heart (specify lesion).—This includes conditions not mentioned in any other categories It might include such lesions as abscess of myocardium or the changes found in the myocardium in vitamin deficiency diseases in thyroid disturbances and those due to parasitic invasion

48 *Rupture of Myocardium (specify location).—This may follow* myocardial infarction or injury of the chest especially if the latter is of the severe crushing type It usually gives rise to precordial pain and may be followed by acute pulmonary edema Rupture into the pericardial sac gives rise to hemopericardium Except in rare instances death follows immediately Rupture of the ventricular septum secondary *to recent infarction gives rise to a systolic thrill and loud harsh* systolic murmur which is loudest along the left border of the sternum in the fourth interspace Rupture of a papillary muscle resulting in marked mitral insufficiency may cause a systolic thrill and a loud often musical murmur in the apical region

49 Thrombosis Within Heart (specify chambers) (See also Mural Thrombosis).—This refers to thrombi occurring within the atrial or ventricular chambers The clinical diagnosis is based upon the occurrence of emboli in the presence of atrial fibrillation mitral stenosis cardiac insufficiency or myocardial infarction (See Pathologic Section) Thrombi in the right chambers embolize to the lungs From the left chambers they embolize to the brain abdominal viscera and the peripheral arteries

In chronic mitral disease with atrial fibrillation large ball like masses (ball valve thrombi) may be found attached to the wall of an atrium or free in the chamber They may occlude the valve orifice and cause sudden attacks of severe cyanosis with dyspnea or of syncope or even sudden death

50 Trauma of Myocardium.—Clinical signs and symptoms vary with the nature of the trauma and the extent and location of the lesion *(i e bullet wound stab wound severe blows on the chest falls or crushing injuries) In contusion of the heart as a result of crushing of* the chest *(e g* steering wheel accident) transient disturbances in rhythm may occur S T displacement and T wave changes in the electrocardiogram serve as evidence of injury to the myocardium The clinical and electrocardiographic findings may simulate those of myo

cardial infarction Hemopericardium is a common sequel of severe injuries of the heart Rupture of the heart may occur but usually is diagnosed only at necropsy

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## DISEASES OF PERICARDIUM

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*Signs*

Faint or absent heart sounds
Increase in area of cardiac dullness
Dullness over sternum replacing normal resonance
Increased venous pressure
Electrocardiogram shows abnormality of S T segment and of T wave
Roentgenological examinations reveal the features mentioned under Serofibrinous Pericarditis

55 HYDROPERICARDIUM —This occurs as part of the anasarca of advanced cardiac insufficiency or *renal failure* or due to neoplastic infiltration The fluid is a transudate The physical signs are as given for serofibrinous pericarditis except that no friction rub is audible

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In chronic mitral disease with atrial fibrillation large ball like masses (ball valve thrombi) may be found attached to the wall of an atrium or free in the chamber They may occlude the valve orifice and cause sudden attacks of severe cyanosis with dyspnea or of syncope or even sudden death

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Roentgenologic examination will show enlarged cardiac silhouette of pyramidal contour with diminished or absent pulsations (Fig. 35 of the Roentgenological Section)

The electrocardiogram shows abnormal S-T segments and T waves as described. If marked effusion occurs the voltage of the QRS complexes may be low

c *Suppurative* – The signs are as in Serofibrinous pericarditis associated with evidences of sepsis. If the purulent effusion takes place rapidly the signs are as in Hemopericardium

3b PERICARDITIS, CHRONIC – The separation into acute and chronic is made arbitrarily on the basis of duration of symptoms and signs. Chronic pericarditis is frequently of rheumatic or tuberculous etiology; other infections and tumors are less frequent causes. The formation of localized adhesions may be a sequel of chronic pericarditis. They may also result from myocardial infarction or injury

a *Adhesive without constriction* – The diagnosis cannot be made unless there are adhesions between the pericardium and mediastinal structures. The following signs are not always reliable diagnostic criteria since they may occur without pericardial adhesions in the presence of much enlarged hearts and in certain other conditions

Signs

Systolic retraction in the region of the apex with diffuse cardiac impulse

Systolic retraction in the eleventh interspace posteriorly on the left side. The sign may occur in thin-chested individuals with great cardiac hypertrophy without adhesions between pericardium and diaphragm

[illegible] on of the apex beat. On turning the patient from one side to the other no shifting of the apex beat is observed

Immobility of the right border of cardiac dullness. On turning the patient from side to side the right border does not move more than approximately 1 cm

Lack of change in the electrical axis of QRS of the electrocardiogram in spite of the above alteration of posture

Roentgenological examination may show enlargement of one or more of the heart chambers. There may be dilatation of the ascending aorta and increased amplitude of pulsations

Electrocardiographic evidence of left ventricular hypertrophy is usual in advanced cases

necropsy Benign cysts are not uncommon and may be diagnosed by roentgenologic technique Occasionally secondary invasion by neoplasm may occur The condition may be suspected if there is a recurrent effusion especially if this be hemorrhagic

57 OTHER STRUCTURAL DISEASES OF PERICARDIUM—This title is to cover diseases not otherwise specified

58 PERICARDITIS ACUTE—a *Fibrinous*—This is often part of the pancarditis of rheumatic fever or of various infectious diseases It may occur at times following upper respiratory infections often believed to be of viral origin It occurs in myocardial infarction if the lesion reaches the epicardial surface and also in uremia It may result from invasion by neoplasm in adjacent tissues

*Symptoms and Signs*

Precordial pain may be present

Pericardial friction rub superficial and harsh usually heard near the left border of the sternum The sound frequently has a to and fro character It may be limited to systole or diastole but may be almost continuous It may last for a few hours or rarely for days

Fever and leucocytosis are often present

The electrocardiogram displays an elevation of the S T junction and segment usually in Leads I and II and in those unipolar extremity and precordial leads which are semidirect leads from the area of the involved pericardium This displacement is temporary and as it disappears the T wave becomes diphasic or inverted in the same leads In certain instances there may be non specific electrocardiographic changes or none at all

b *Serofibrinous*—The etiology of this type is usually the same as for the fibrinous type An effusion exudative in nature and variable in amount occurs and may be sanguineous

*Signs*

Paradoxical pulse

Dullness over the sternum replacing normal resonance

Dullness and bronchial breathing in left infrascapular area

Faint heart sounds

*Pericardial friction* rub usually limited to the basal portion of the heart

Roentgenologic examination will show enlarged cardiac silhouette of pyramidal contour with diminished or absent pulsations (Fig. 35 of the Roentgenological Section)

The electrocardiogram shows abnormal ST segments and T waves as described. If marked effusion occurs the voltage of the QRS complexes may be low.

c. *Suppurative*.—The signs are as in Serofibrinous pericarditis associated with evidences of sepsis. If the purulent effusion takes place rapidly the signs are as in Hemopericardium.

49. PERICARDITIS, CHRONIC.—The separation into acute and chronic is made arbitrarily on the basis of duration of symptoms and signs. Chronic pericarditis is frequently of rheumatic or tuberculous etiology; other infections and tumors are less frequent causes. The formation of localized adhesions may be a sequel of chronic pericarditis. They may also result from myocardial infarction or injury.

a. *Adhesive without constriction*.—The diagnosis cannot be made unless there are adhesions between the pericardium and mediastinal structures. The following signs are not always reliable diagnostically since they may occur without pericardial adhesions in the presence of much enlarged hearts and in certain other conditions.

Signs

- Systolic retraction in the region of the apex with diffuse cardiac impulse.
- Systolic retraction in the eleventh interspace posteriorly on the left side. The sign may occur in thin chested individuals with great cardiac hypertrophy without adhesions between pericardium and diaphragm.
- Fixation of the apex beat. On turning the patient from one side to the other no shifting of the apex beat is observed.
- Immobility of the right border of cardiac dullness. On turning the patient from side to side the right border does not move more than approximately 1 cm.
- Lack of change in the electrical axis of QRS of the electrocardiogram in spite of the above alteration of posture.
- Roentgenological examination may show enlargement of one or more of the heart chambers. There may be dilatation of the ascending aorta and increased amplitude of pulsations.
- Electrocardiographic evidence of left ventricular hypertrophy is usual in advanced cases.

b *Constrictive*—This is frequently of unknown etiology but may result from tuberculosis or septic infection or may be part of a polyserositis The clinical picture may vary with the site of the constriction whether affecting chiefly the atria the ventricles or the venous orifices The heart may be enlarged but is often normal or even small Occasionally the adhesions become calcified

*Signs*

Increased venous pressure
Low arterial pressure with small pulse pressure
Roentgenologic examination usually reveals normal or small heart
Small or absent pulsations of heart shown by fluoroscopy or kymography
Calcification of the pericardium may be demonstrable (Figs 38 39 of the Roentgenological Section)
The electrocardiogram usually shows low voltage of the QRS complexes and low voltage or inversion of T waves
Recurrent ascites
Enlarged liver

c *Serous*—Tuberculosis is probably the commonest cause of this form of pericarditis The amount of effusion is usually considerably greater than in the serofibrinous form The signs are as in Serofibrinous pericarditis but a pericardial friction rub is usually lacking

60 PNEUMOPERICARDIUM—This is most often a result of injury and is usually complicated by serous suppurative or sanguinous effusion Tympany replaces the cardiac dullness and if fluid is present splashing or churning sounds may be heard On roentgenologic examination the air within the pericardium is apparent

61 TRAUMA TO PERICARDIUM (specify character of lesion)—Bullet or stab wounds may involve the pericardium at times without injury to the myocardium Foreign bodies may also reach the pericardial sac through the esophagus or the bronchi and give rise to suppurative pericarditis Occasionally hemopericardium occurs

The signs and symptoms depend upon the character and extent of the injury and the occurrence of complications *e g* hemorrhage or inflammation (See Hemopericardium and Pericarditis Acute (c *Suppurative*) )

# PHYSIOLOGICAL DIAGNOSIS

THE PHYSIOLOGICAL diagnosis covers three main categories of cardiac physiology, the cardiac mechanism, valvular incompetence and the clinical syndrome which may result from the patient's disease. A title from each category should be mentioned if indicated. The importance of the designation of the clinical syndrome lies in giving a picture of the clinical aspects of the case.

## CARDIAC MECHANISM

The diagnosis of irregularities of the heart and other disturbances of the mechanisms of the heart can frequently be made by physical examination alone. At times the use of instrumental methods may be necessary to arrive at a diagnosis.

1. ARRHYTHMIA (undiagnosed).—This caption should be used for those arrhythmias which cannot be diagnosed clinically or instrumentally. It is also to be used when a complete examination is not possible and the nature of the arrhythmia is in doubt or when there is only a history of arrhythmia.

2. ATRIAL FIBRILLATION.—This is an incoordinated irregular beating of the atria, the major muscular movements usually occurring at a rate between 350 and 500 per minute. It may be attributed to a circus movement within the atrial muscle which does not follow a constant path.* The ventricles respond irregularly, the rate depending in large measure upon the facility of atrioventricular conduction. The ventricular rate without treatment is generally between 90 and 160 per minute. Occasionally rates as low as 40 to 60 per minute may be observed, especially when the heart is under the influence of digitalis. When the ventricular rate is rapid or slow the irregularity may escape detection. When complete heart block is present the ventricular rhythm may be regular.

Atrial fibrillation should be suspected when the heart beat is grossly

* It should be stated that this explanation has been questioned. It has been suggested that atrial flutter and fibrillation are of the same genus as paroxysmal tachycardia, the rate of the stimulus being the determining factor. It is most rapid in atrial fibrillation, less rapid in flutter and least rapid in tachycardia.

irregular Often there are varying periods of rapid and slow beats Exercise increases the heart rate and accentuates the irregularity whereas the *arrhythmia due to premature contractions usually disappears* with an increase of heart rate The electrocardiogram will confirm the diagnosis

Atrial fibrillation may be (a) Transient (paroxysmal) or (b) Persistent (chronic)

3 ATRIAL FLUTTER —This is a rapid coordinated contraction of the atrial muscle usually occurring at a rate between 200 and 350 per minute It may be attributed to a circus movement within the atrial muscle following a constant path The ventricular rate is commonly one half but may be one third or one fourth of the atrial rate depending upon the facility of A V conduction The ventricular rhythm usually is regular but may be irregular or may change from one to the other

Atrial flutter should be suspected when the ventricular rate is about 150 per minute and the rhythm regular Under these conditions the *rate is unaffected by change in position by rest or by moderate exercise* There is generally marked temporary slowing of the ventricles during carotid sinus pressure When this occurs the rhythm may become irregular The electrocardiogram will usually confirm the diagnosis

*Atrial flutter* may be (a) Transient (paroxysmal) or (b) *Persistent* (chronic)

4 ATRIOVENTRICULAR BLOCK —The conduction of the impulses from atria to ventricles is impaired

a *Incomplete A V block* (prolonged conduction time) —This can be detected only by a graphic record The record shows the atrioventricular conduction time beyond the normal limit of 0 2 second

b *Incomplete A V block* with dropped beats indicates a greater impairment of conduction Dropped beats due to heart block may be suspected when *auscultation reveals a regular rhythm interrupted by* pauses apparently double the normal A dropped beat due to incomplete heart block may be confused with the long pause which follows an early premature ventricular contraction They may be distinguished by the absence of any sound during the long pause in heart block and by *the presence of the* sound of the premature beat following shortly after the second sound of the preceding normal beat when the arrhythmia is due to premature contractions A graphic record will establish the diagnosis Dropped beats due to sinus arrest to blocked atrial premature *systoles and to incomplete* heart block can be differentiated best by the electrocardiogram

c. *Complete A V block*.—This should be suspected when the ventricular rate is under 40 per minute and the rhythm is regular. Atrial sounds may be heard occasionally in diastole or sometimes accentuating the first heart sound. In the jugular pulse and especially if a tracing is made, atrial waves may be observed between the carotid pulsations. The ventricular rate varies little or not at all with change of position or after exercise. There may be prolonged periods of ventricular standstill or of ventricular fibrillation during which syncope and convulsions may occur (Adams Stokes Syndrome). A graphic record confirms the diagnosis.

5. ATRIOVENTRICULAR NODAL RHYTHM.—This rhythm originates in the atrioventricular node, usually occurring when the sino-atrial node is depressed. The rate is generally between 65 and 85 per minute but is subject to wide variations. The diagnosis can be made only from an electrocardiogram.

6. ESCAPED BEATS (Ventricular Escape).—The ventricles may beat from an intrinsic stimulus when the impulse initiated at the sinus node is delayed or is prevented from reaching the ventricles. This independent beat occurs after a long pause and usually originates in the atrioventricular node or the bundle of His. If it arises in the A V node it is called *A V nodal escape*; if in the bundle of His, *His bundle escape*. If it arises below the bifurcation of the bundle it is properly termed *ventricular escape*. The term however is often used to describe the phenomenon regardless of the focus of origin.

7. INTRAVENTRICULAR BLOCK.—Conduction is defective below the bifurcation of the atrioventricular bundle. Impaired conduction may manifest itself in either the right or left bundle branch or in any subdivision of the intraventricular conduction system. Diagnosis can be made only by means of an electrocardiogram.

8. OTHER ARRHYTHMIAS (specify).—Under this heading may be filed conditions not included in the categories here given.

9. PAROXYSMAL TACHYCARDIA is a rapid and regular succession of ectopic beats which occur in paroxysms of varying duration. The rate is usually more than 120 and may be as much as 270 per minute. It usually exceeds 150 per minute. The beginning and ending of the attacks are usually abrupt and the onset is generally recognized by the patient.

Paroxysmal tachycardia may be suspected when there is a rapid regular rhythm which is unaffected by position, rest or exercise. Carotid sinus pressure may terminate the attack if the origin is in the atria or the atrioventricular node but otherwise will not slow the rate.

The paroxysms originate at various points outside of the sinus node. The origin may be

a. Atrial
b. Atrioventricular nodal
c. Unknown supraventricular origin
d. Ventricular

Differential diagnosis of the site of origin can be made with certainty only from the electrocardiogram.

10. PREMATURE CONTRACTIONS.—The heart's rhythm is interrupted by a contraction originating prematurely in a focus outside of the sinus node. A premature contraction rarely may arise in the sinus node itself. This interruption may occur regularly or irregularly. When a premature contraction interrupts a regular sinus rhythm, it is recognized on auscultation as a beat occurring before the expected time of the next normal beat. It is followed by a longer pause than normal. These characteristics are often obscured when a premature contraction occurs during an irregular rhythm or when it is completed without interfering with the next beat of the basic rhythm (interpolated premature beat).

On palpation of the radial artery a premature beat or an intermission of the pulse will be found to correspond with the premature heart sounds.

Exercise, by increasing the heart rate, will often abolish premature contractions, whereas an irregularity due to atrial fibrillation will persist. When the rate slows after exercise an irregularity due to premature contractions may become exaggerated. Occasionally exercise will result in an immediate increase in the number of premature contractions.

The point of origin of premature contractions can be determined with accuracy only from the electrocardiogram, though a ventricular origin is by far the most frequent.

a. Atrial
b. Atrioventricular nodal
c. Unknown supraventricular origin
d. Ventricular

11. PULSUS ALTERNANS.—The rhythm is regular but the arterial pulses alternate in size. This may be detected by palpation of the

pulse It is best observed while the blood pressure is taken by the auscultatory method The sounds then appear to be evenly spaced but show a regular alternation in intensity Near the upper level of systolic pressure the feeble sounds may be inaudible

Pulsus Alternans must be distinguished from the regular occurrence of premature contractions for which purpose a graphic record may be needed

12 SINUS ARREST (sino atrial block) —Standstill of the entire heart for a period of one cycle or longer may occur because the sinus node is quiescent or because the impulse coming from it is blocked before reaching the atria In the course of a sinus rhythm a pause occurs which is longer than the duration of one cycle Sinus arrest must be distinguished from the pause associated with an early premature contraction or from heart block with dropped beats Clinically it may be suspected on noticing what appears to be a dropped beat by auscultation but the true character can be recognized only from the electrocardiogram Under certain conditions much longer pauses may occur

13 SINUS ARRHYTHMIA is due to an irregularity of impulse formation in the sinus node It should be suspected when the rate slows progressively for a short series of beats and the slowing is followed by a gradual quickening This variation is often persistent Ordinarily quickening occurs with inspiration slowing with expiration At times the phases of sinus arrhythmia do not coincide with the phases of respiration It is common in children and in the aged An electrocardiographic record will confirm the diagnosis

14 SINUS BRADYCARDIA —This is a slow sinus rhythm with a rate less than 60 per minute which tends to become slower when the patient is recumbent and more rapid when standing and after exercise

15 SINUS RHYTHM NORMAL—This is the normal rhythm of the heart originating in the sinus node The rate is between 60 and 100 per minute at rest The rate tends to become slower when the patient is recumbent and more rapid when standing and after exercise

16 SINUS TACHYCARDIA —This is a rapid sinus rhythm with a rate of more than 100 per minute The rate tends to become slower when the patient is recumbent and more rapid when standing and after exercise

17 VENTRICULAR FIBRILLATION —This is a rapid irregular mechanically ineffectual contraction of the ventricular muscle It is usually

terminal event though transient attacks may occur It is recognized only from the electrocardiogram

18 WANDERING PACEMAKER —*The site of the stimulus may change* its location within the sinus node or may shift to the A V node An arrhythmia usually results which often resembles sinus arrhythmia It is recognized only by the electrocardiogram

## VALVULAR INCOMPETENCE

19 VALVULAR INCOMPETENCE —Valvular incompetence is due to dilatation of the valve ring It is to be distinguished from valvular deformity with insufficiency in that there is no structural alteration of the valve leaflet *It is considered a physiological rather than an anatomical* diagnosis because the dilatation is primarily the result of a disturbance of cardiac function and may be reversible not indicating permanent disturbance of the structure of the valve

a *Aortic Incompetence* —A diastolic murmur like that of aortic insufficiency may occur in the absence of aortic valvular deformity

b *Mitral Incompetence* —Evidence of mitral incompetence is af*forded by a systolic murmur which has the same characteristic as the* murmur described for insufficiency due to valvular deformity The differentiation is made largely by inference keeping in mind the following features (1) Mitral incompetence occurs commonly in cardiac insufficiency and especially if marked enlargement is present (2) When there is disease of the myocardium or aortic valve without rheumatic etiology the murmur is probably due to incompetence and not to valvular disease (3) When there is a rheumatic history with aortic valvular disease it may be impossible to decide whether the murmur is produced by deformity of the mitral valve or by incompetence Without marked enlargement or cardiac insufficiency valvular deformity is the more likely cause (4) In the presence of mitral stenosis the murmur must be due to valvular deformity and therefore should be diagnosed as mitral insufficiency (5) At times the murmur must be differentiated from innocent (unexplained) systolic murmurs heard at the apex

c *Pulmonic incompetence* —The murmur of pulmonic incompetence cannot be indistinguished by its quality from that due to pulmonic insufficiency but it is typically a transitory phenomenon It must be distinguished from the murmur of aortic insufficiency It often is heard in association with pulmonary arterial hypertension such as found with marked mitral stenosis and cardiac insufficiency and is then called the Graham *Steell murmur*

d *Tricuspid incompetence* —The signs are not distinguishable from those described for tricuspid insufficiency due to valvular deformity The diagnosis as in mitral incompetence is made largely by inference Tricuspid incompetence is secondary to preexisting mitral deformity or incompetence or to pulmonary arterial hypertension

## CLINICAL SYNDROMES

These are physiological disturbances originating from cardiac dysfunction which have clinical manifestations distinctive enough to merit inclusion in the diagnosis

20 Adams Stokes Syndrome —This term is applied to attacks characterized by unconsciousness often accompanied by muscular twitchings and even general convulsions They occur in patients with atrioventricular block when there is ventricular asystole or fibrillation for a sufficient period of time to result in a severe grade of cerebral ischemia The duration and severity of the attack depend upon the duration of the asystole The term is not to be applied to syncope due to other causes

21 Anginal Syndrome —In this syndrome the major symptom is thoracic pain which is precipitated usually by effort but sometimes by excitement a heavy meal or exposure to cold The pain is usually substernal or just to the left of the sternum Occasionally the pain is epigastric and in rare instances it may be localized in the neck or the left arm or shoulder

There is a tendency for the pain to radiate most frequently to the left shoulder and arm and occasionally to the fingers Less frequently it may radiate to the neck jaw and teeth to the back upper abdomen or to the right shoulder and arm At times the pain will start at one of these points before focusing on the anterior surface of the chest

The intensity varies from a slight sense of heaviness to a severe crushing pain Since the precipitating cause is commonly physical exertion rest usually causes the pain to subside The length of the episode therefore is relatively short Occasionally an attack may come on while the patient is at rest or even when asleep

The pain is often accompanied by a sense of choking or inability to breathe which is also relieved by rest The patient will often complain of flatulence as well If the attack is not relieved by rest or a nitrite and lasts for an hour or more and especially if it is accompanied by circulatory collapse myocardial infarction should be strongly sus

pected Occasionally the pain of myocardial infarction may be identical with the pain of the anginal syndrome The associated symptomatology and the subsequent course will determine the diagnosis

Anginal syndrome is a physiological diagnosis and is not synonymous with coronary arteriosclerosis The underlying mechanism is hypoxia of the heart muscle so that any factor organic or functional which may invoke such a mechanism can initiate the anginal syndrome Neurocirculatory asthenia may produce symptoms simulating the anginal syndrome but a careful history should make the distinction possible

22 Cardiac Insufficiency (heart failure) occurs when the heart fails to circulate the optimal amount of blood required by the body Early in the process the clinical picture may be predominantly that of insufficiency of the one ventricle which has primarily failed Later the clinical picture is generally that of failure of both ventricles in varying degrees In cardiac insufficiency the cardiac output (minute volume) is always less than required There is an inadequate emptying of the cardiac chambers increased diastolic pressures therein and a greater than normal deoxygenation of the blood (increased arteriovenous oxygen difference)

In the initial phases of its development cardiac insufficiency manifests itself only when the demand upon the circulation is increased as with exercise It may not be present during the lesser circulatory requirements of the resting state With further development of cardiac insufficiency the ability of the heart to meet increased demands upon the circulation is progressively diminished The different degrees of this ability are to be expressed by the grades of Cardiac Functional Capacity

The failure of the ventricles to circulate the required volume of blood leads to congestion on the venous side of the vascular tree either pulmonary or systemic or both (backward failure congestive failure) and to inadequate blood flow on the arterial side (forward failure) with resulting alterations in the functions of the organs As a result of these processes the circulation time may be prolonged Renal function often is affected causing retention of salt and water and usually an increase of blood volume which further aggravates the congestive failure

The symptoms and signs of cardiac insufficiency depend upon which ventricle fails primarily and on the degree and duration of its failure Dyspnea fatigue and cyanosis are the cardinal symptoms

*Insufficiency of the Left Ventricle* results in stasis in the pulmonary circuit with dyspnea and orthopnea pulmonary rales and diminished

vital capacity Acutely developing left ventricular failure causes the syndrome of *Paroxysmal Dyspnea* This appears quite suddenly usually at night with tachypnea severe dyspnea and orthopnea Examination of the lungs reveals prolonged expiration often wheezing with or without rales A more severe stage of the same process is described as *Paroxysmal Pulmonary Edema* There is severe respiratory distress cyanosis and cough with profuse frothy and perhaps pinkish sputum Large and small rales and ronchi are heard throughout the lungs

*Insufficiency of the Right Ventricle* results in stasis in the systemic venous circuit The veins of the neck and arms show an increased pressure the liver is enlarged and is tender if distension has been acute There may be edema of the dependent parts of the body hydrothorax and ascites

23 CAROTID SINUS SYNDROME —Over irritability of the carotid sinus may cause attacks of dizziness fainting and sometimes convulsions During the attack there is usually a fall in arterial blood pressure and a marked slowing of the heart rate Attacks may come on without any apparent cause they may follow an emotional upset they may be produced by pressure over the carotid sinus or by swallowing

pected. Occasionally the pain of myocardial infarction may be identical with the pain of the anginal syndrome. The associated symptomatology and the subsequent course will determine the diagnosis.

Anginal syndrome is a physiological diagnosis and is not synonymous with coronary arteriosclerosis. The underlying mechanism is hypoxia of the heart muscle so that any factor, organic or functional, which may invoke such a mechanism can initiate the anginal syndrome. Neurocirculatory asthenia may produce symptoms simulating the anginal syndrome but a careful history should make the distinction possible.

22. Cardiac Insufficiency (heart failure) occurs when the heart fails to circulate the optimal amount of blood required by the body. Early in the process the clinical picture may be predominantly that of insufficiency of the one ventricle which has primarily failed. Later the clinical picture is generally that of failure of both ventricles in varying degrees. In cardiac insufficiency the cardiac output (minute volume) is always less than required. There is an inadequate emptying of the cardiac chambers, increased diastolic pressures therein and a greater than normal deoxygenation of the blood (increased arteriovenous oxygen difference).

In the initial phases of its development cardiac insufficiency manifests itself only when the demand upon the circulation is increased, as with exercise. It may not be present during the lesser circulatory requirements of the resting state. With further development of cardiac insufficiency the ability of the heart to meet increased demands upon the circulation is progressively diminished. The different degrees of this ability are to be expressed by the grades of Cardiac Functional Capacity.

The failure of the ventricles to circulate the required volume of blood leads to congestion on the venous side of the vascular tree, either *pulmonary or systemic or both (backward failure, congestive failure)*, and to inadequate blood flow on the arterial side (forward failure) with resulting alterations in the functions of the organs. As a result of these processes the circulation time may be prolonged. Renal function often is affected causing retention of salt and water and usually an increase of blood volume which further aggravates the congestive failure.

The symptoms and signs of cardiac insufficiency depend upon which ventricle fails primarily and on the degree and duration of its failure. Dyspnea, fatigue and cyanosis are the cardinal symptoms.

*Insufficiency of the Left Ventricle* results in stasis in the pulmonary circuit with dyspnea and orthopnea, pulmonary rales, and diminished

history particularly with reference to the patient's symptoms on effort An accurate account of the reaction produced by varying degrees of exertion—such as walking on the level or up a grade ascending stairs or running—is an essential part of the history If there is difficulty in deciding on the proper rating by this method direct observation of a patient during and after the performance of a moderately strenuous exercise may be helpful The occurrence of undue dyspnea or exhaustion or cardiac pain is of special significance

In general the more intense the subjective symptoms the more likely that there will be physical signs of cardiac disease Discrepancies often exist between the number and intensity of the physical signs and the degree of the subjective distress on effort This is apt to be true particularly in patients suffering from the anginal syndrome in whom objective evidences of disease may be slight or absent With exertional dyspnea special care should be taken to rule out or to evaluate the role of the pulmonary causes of dyspnea such as obstructive emphysema bronchial asthma and pulmonary fibrosis

The classification of patients according to their cardiac functional capacity gives only a part of the information needed to plan the management of the patients activities A diagnosis to indicate the regulation of physical activity should be based on information derived from many sources and is considered in the section on Therapeutic Classification The functional classification is an estimate of what the patient's heart will allow him to do and should not be influenced by the character of the structural lesions or by an opinion as to treatment or prognosis

## Classification of Patients

CLASS I Patients with cardiac disease but without resulting limitation of physical activity Ordinary physical activity does not cause undue fatigue palpitation dyspnea or anginal pain

CLASS II Patients with cardiac disease resulting in slight limitation of physical activity They are comfortable at rest Ordinary physical activity results in fatigue palpitation dyspnea or anginal pain

CLASS III Patients with cardiac disease resulting in marked limitation of physical activity They are comfortable at rest Less than ordinary activity causes fatigue palpitation dyspnea or anginal pain

CLASS IV Patients with cardiac disease resulting in inability to carry on any physical activity without discomfort Symptoms of cardiac insufficiency or of the anginal syndrome may be present even at rest If any physical activity is undertaken discomfort is increased

# FUNCTIONAL CAPACITY

At the present time there is no clinical test which will measure accurately the functional capacity of the heart For the purposes of this classification it is to be estimated by appraising the patient s ability to perform physical activity The estimate is only approximate for it is derived largely by inference from the history It represents an expression of opinion concerning the functional capacity of the patient as modified specifically by his cardiac disease Usually structural changes are present in the heart Occasionally as for instance in certain cases of atrial fibrillation or paroxysmal tachycardia or in certain patients suffering from the anginal syndrome no anatomical lesions can be detected

The *diminution in functional capacity which results from a cardiac* disease may be accompanied by discomfort or by signs of impaired circulation or both The extent to which physical activity is curtailed thereby and the severity of the symptoms caused by effort are used in estimating the degree of reduction of functional capacity Functional capacity *often is limited because of cardiac insufficiency Physical* signs then may be present or absent but their presence or absence should not influence the rating

In the estimation of cardiac functional capacity the term *ordinary physical activity* is used to describe the actual physical performance of which each patient was capable prior to the onset of manifest cardiac disease or such activity as would be normal for an individual of the same age sex and physical development In case of cardiac disease dating from early childhood a patient s normal functional capacity can only be estimated by such a comparison The presence of active disease in the heart acute infectious diseases convalescence muscular weakness anemia arthritis pulmonary disease chest deformity obesity and psychogenic disability may interfere with judging the cardiac functional capacity by imposing limitations from these causes For example functional capacity is not to be regarded as limited even though organic heart disease be present when it is *clear that the patient s* incapacity is due to a psychoneurotic state

In estimating a patient s response to effort a comparison must be made between his ordinary physical activity and his present capacity for physical exertion Usually this estimate may be based upon the

## NO HEART DISEASE PREDISPOSING ETIOLOGICAL FACTOR*

These are patients in whom no cardiac disease is discovered but whose course should be followed by periodic examinations because of the presence or history of an etiological factor which might cause heart disease. These cases should be recorded as No Heart Disease, Predisposing Etiological Factor and it is essential that the etiological diagnosis should also be stated.

## UNDIAGNOSED MANIFESTATION*

Patients with symptoms or signs referable to the heart but in whom a diagnosis of cardiac disease is uncertain should be classified tentatively as Undiagnosed Manifestation.

Reexamination after a suitable interval will usually help to establish a definite diagnosis. When there is a reasonable probability that the signs or symptoms are not of cardiac origin the title Undiagnosed Manifestation should not be used. The diagnosis then should be No Heart Disease.

* There are patients in whom the *symptoms* or signs, though suggestive of cardiac disease, do not justify a definite diagnosis and from whom is obtained a history of an etiological factor which might cause heart disease. The diagnosis in such cases should include both No Heart Disease, Predisposing Etiological Factor and Undiagnosed Manifestation.

## THERAPEUTIC CLASSIFICATION

The Therapeutic Classification is intended as a guide to the management of cardiac patients. It gives a prescription for the amount of physical activity which is advised for those in each class. The functional capacity of the patient does not always determine the amount of physical activity which should be permitted. For example, a child with active rheumatic carditis may not experience discomfort on playing baseball, yet rest in bed is imperative. Such a patient would be designated as Class I (Functional) E (Therapeutic). There is frequently a difference between the amount of physical activity which the patient can undertake in terms of his functional capacity and that which he should attempt in order to prevent aggravation of the disease. The recommendation as to physical activity is based not only upon the *amount of effort possible without discomfort but also upon the nature* and severity of the cardiac disorder.

### Classification of Patients

CLASS A. Patients with cardiac disease whose physical activity need not be restricted.

CLASS B. Patients with cardiac disease whose ordinary physical activity need not be restricted, but who should be advised against severe or competitive physical efforts.

CLASS C. Patients with cardiac disease whose ordinary physical activity should be moderately *restricted, and whose more strenuous efforts* should be discontinued.

CLASS D. Patients with cardiac disease whose ordinary physical activity should be markedly restricted.

CLASS E. Patients with cardiac disease who should be at complete rest, confined *to bed or chair*.

# GUIDE TO THE ROENTGENOLOGICAL DIAGNOSIS

# NOMENCLATURE

1 Normal Heart
   a Horizontal
   b Vertical
   c. Oblique or Globular

2 Left Ventricle
   a Enlargement in Length Outflow Tract
   b Enlargement in Width and Depth Inflow Tract
   c. Concentric Hypertrophy
   d Localized Ventricular Bulge (Ventricular Aneurysm)
   e Localized Impairment of Contraction

3 Right Ventricle
   a Enlargement in Length Outflow Tract
   b Rotation of Heart on its Vertical Axis
   c. Enlargement in Width and Depth Inflow Tract

4 Left Atrium
   a Posterior or Horizontal Enlargement
   b Upward or Vertical Enlargement
   c. Enlargement toward Right
   d Enlargement toward Left

5 Right Atrium
   a Enlargement of Auricle (Auricular Appendix)
   b Enlargement Posteriorly

6 Symmetrical Cardiac Enlargement

7 Displacement of Heart

8 Calcification Intracardiac
   a Valvular
   b Annulus Fibrosus
   c Coronary Arteries
   d Myocardium
   e Endocardium

9 Pericardium
   a Effusion
   b Adhesions
   c Constriction
   d Calcification
   e Cysts

## TECHNIQUE

ROENTGENOLOGIC examination of the heart enables the examiner to observe the contours the size and position of the heart and that of its individual chambers The usual methods of roentgenologic examinations are

1 Roentgenoscopy (Fluoroscopy)
2 Teleoroentgenography
3 Orthodiagraphy

Special methods Roentgenkymography
Angiocardiography

1 *Roentgenoscopy (Fluoroscopy)* is the direct examination of the patient behind the fluorescent screen The closeness of the object to the target of the roentgen tube results in an exaggeration in size of the objects viewed so that this technique is unsatisfactory for measurements To determine exactly the size of the cardiac shadow teleoroentgenography or orthodiagraphy must be used The cardiac contour should be studied fluoroscopically not only in the postero-anterior position but also in various degrees of rotation so that depth also may be estimated The patient should be turned sufficiently so that the border to be examined appears clearly and free of the shadow of the vertebrae The amount of rotation needed to effect this is usually about fifty degrees to the right or left but may be more or less depending upon whether the chest approaches the hypersthenic or the hyposthenic type upon the presence of emphysema scoliosis or kyphosis and upon the degree of enlargement of the cardiac chambers Abduction of the arms or elevation of the scapula may be used to clear the field of vision The standard positions are (Figs 1 2 3)

Postero-Anterior Patient facing the screen

Right Anterior Oblique Patient facing the screen but turned so that the right shoulder is toward the screen

Left Anterior Oblique Patient facing the screen but turned so that the left shoulder is toward the screen

The normal cardiac configuration may be altered by displacement of the heart or by enlargement of one or more of its chambers By the roentgenoscopic method it is possible to observe the outline of the individual chambers of the heart In the oblique positions the esophagus and the bifurcation of the trachea serve as aids in determin

10 AORTA
- a Elongation and Dilatation
- b Calcification
- c Hypoplasia

11 ANEURYSM OF AORTA
- a Saccular
- b Fusiform
- c Dissecting Hematoma

12 BRACHIOCEPHALIC ARTERIES, WIDENING OF

13 SUPERIOR VENA CAVA DILATATION OF

14 PULMONARY ARTERY
- a Dilatation of Trunk and Primary Branches
- b Dilatation of Secondary or Hilar Vessels
- c Narrowing of Pulmonary Arteries

15 PULMONARY FIELDS
- a Hilar Dilatation
- b Pulmonary Stasis Acute Chronic Localized Diffuse
- c Pleural Effusion Thickening

Measurements greater than the predicted normal are sometimes obtained in normal subjects and normal measurements are sometimes obtained from hearts with evident enlargement of individual chambers Indeed a measurement ten per cent greater than the prediction figure may occasionally be obtained from a normal heart This lessens the value of the measurement in the borderline group where a decisive answer would be most desirable but in spite of this fact it is the best available method of measurement There still may be borderline cases in which neither measurement nor fluoroscopic study of the contours will answer the question as to whether or not enlargement is present

The chief use of measurements is for the comparison of serial roentgenograms in the individual case In advanced cardiac enlargement the exact estimation of size is useful in following the process It may however be more important to ascertain which chambers participate in the enlargement and to what degree Such information cannot be obtained by measurement and is probably best obtained by fluoroscopy

3 *Orthodiagraphy* utilizes central parallel rays to outline the cardiac contour enabling the examiner to make a drawing closely approximating in size and shape the outline of the heart in the plane under observation The outlines of the diaphragm and of the median line are similarly drawn The lower contour of the heart and the upper contour at the base should not be attempted as the opacity of the liver and of the vertebrae interfere The transverse diameter or the surface area is compared with prediction formulas based upon age height and weight to determine the presence of enlargement (Table II) The orthodiagraphic method is subject to but slight error in skilled hands The interpretation of the tracing is subject to the same difficulties that have been discussed under Teleoroentgenography

*Roentgenkymography* is the graphic registration of the pulsations of the contours of the heart This may be accomplished by means of roentgen rays directed at a moving film in contact with a stationary grid having vertical or horizontal slits (conventional method) or by the use of a photoelectric cell amplification and by appropriate shielding and recording devices (roentgenelectrokymography)

*Angiocardiography* is the method of visualizing the heart chambers and great vessels by the rapid intravenous injection of a radiopaque substance Serial roentgenograms are taken at appropriate intervals

ing enlargement of certain chambers as do also the character and timing of the pulsations of the individual chambers and of the great vessels and the localization of the interventricular septum and groove Examination in oblique views may reveal enlargement of one or more chambers when the usual postero anterior view shows a normal configuration

It requires considerable experience to judge the range of normal variation The best procedure is to combine roentgenoscopic description of the individual heart chambers with teleoroentgenograms preferably including oblique views though orthodiagraphy in the same positions may serve as well The value of permanent records for purposes of comparison is obvious

2 *Teleoroentgenography* is the taking of chest roentgenograms with the tube at a standard distance usually two meters from the film At such distance the divergent rays which delineate the borders of the cardiac shadow exaggerate its size by from four to twelve per cent For the P A view the patient s sternum is in contact with the film cassette The target of the roentgen tube is centered at the level of the sixth thoracic vertebra Synchronization of the exposure with systole or diastole may at times make a significant difference but the most important feature in the determination of heart size is that the picture must be made with the diaphragm in a standard position Exposures should be made *at the end of a normal inspiration* as otherwise standards cannot be applied

Roentgenograms taken in the right and left oblique positions are desirable and are recommended to supplement the P A view and the roentgenoscopic findings Lateral views have the value of being taken in a position that is reproduceable for serial records but do not give as much information as to the size and shape of the different cardiac chambers and the aorta as do the oblique views

For measurement of the P A view a vertical line is drawn approximately through the spinous processes of the lower dorsal vertebrae The perpendiculars from this line to the most distant point of the right and left borders are added together to make the *Transverse Diameter* (T D ) The ratio between the transverse diameter of the heart and the transverse diameter of the thorax (*the cardio thoracic ratio*) *should be disregarded* as it is an unreliable index of cardiac enlargement

Tables of normal cardiac measurements and their variation within physiological limits as affected by height weight and age are found on later pages The disadvantage of prediction formulas applied to teleoroentgenography and to orthodiagraphy is that they do not always give a correct separation between hearts of normal and abnormal size

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TABLE I PREDICTION TABLE FOR AVERAGE NORMAL TRANSVERSE DIAMETERS OF THE HEART SILHOUETTE BASED ON THE TELEROENTGENOGRAM

| T. D. of Heart | HEIGHT | | | | | | | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | 5 0 | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8" | 9 | 10 | 11 | 6 0 | 1 | 2 | 3 | 4 | 5 | 6 |
| 100 mm | 83 | 85 | 86 | 87 | 89 | 90 | 92 | | | | | | | | | | | | |
| 101 | 85 | 86 | 88 | 89 | 91 | 92 | 93 | 95 | | | | | | | | | | | |
| 102 | 87 | 88 | 90 | 91 | 92 | 94 | 95 | 97 | | | | | | | | | | | |
| 103 | 88 | 90 | 92 | 93 | 94 | 96 | 97 | 99 | 100 | | | | | | | | | | |
| 104 | 90 | 92 | 93 | 95 | 96 | 98 | 99 | 101 | 102 | | | | | | | | | | |
| 105 | 92 | 93 | 95 | 96 | 98 | 99 | 101 | 103 | 104 | 106 | | | | | | | | | |
| 106 | 94 | 95 | 97 | 98 | 100 | 101 | 103 | 104 | 106 | 108 | | | | | | | | | |
| 107 | 95 | 97 | 99 | 100 | 102 | 103 | 105 | 106 | 108 | 110 | 111 | | | | | | | | |
| 108 | 97 | 99 | 100 | 102 | 104 | 105 | 107 | 108 | 110 | 112 | 113 | | | | | | | | |
| 109 | 99 | 101 | 102 | 104 | 106 | 107 | 109 | 110 | 112 | 114 | 115 | 117 | | | | | | | |
| 110 | 101 | 102 | 104 | 106 | 108 | 109 | 111 | 113 | 114 | 116 | 118 | 119 | 121 | | | | | | |
| 111 | 103 | 104 | 106 | 108 | 109 | 111 | 113 | 115 | 116 | 118 | 120 | 121 | 123 | 125 | | | | | |
| 112 | 105 | 106 | 108 | 110 | 111 | 113 | 115 | 117 | 118 | 120 | 122 | 124 | 125 | 127 | 129 | | | | |
| 113 | 106 | 108 | 110 | 112 | 113 | 115 | 117 | 119 | 121 | 123 | 124 | 126 | 128 | 129 | 131 | 133 | | | |
| 114 | 108 | 110 | 112 | 114 | 115 | 117 | 119 | 121 | 123 | 125 | 126 | 128 | 130 | 132 | 133 | 135 | 137 | | |
| 115 | 110 | 112 | 114 | 116 | 117 | 119 | 121 | 123 | 125 | 127 | 129 | 130 | 132 | 134 | 136 | 138 | 140 | 141 | |
| 116 | 112 | 114 | 116 | 118 | 120 | 121 | 123 | 125 | 127 | 129 | 131 | 133 | 134 | 136 | 138 | 140 | 142 | 144 | 146 |
| 117 | 114 | 116 | 118 | 120 | 122 | 124 | 125 | 127 | 129 | 131 | 133 | 135 | 137 | 139 | 141 | 143 | 144 | 146 | 148 |
| 118 | 116 | 118 | 120 | 122 | 124 | 126 | 128 | 129 | 131 | 133 | 135 | 137 | 139 | 141 | 143 | 145 | 147 | 149 | 151 |
| 119 | 118 | 120 | 122 | 124 | 126 | 128 | 130 | 132 | 134 | 136 | 138 | 140 | 142 | 143 | 145 | 147 | 149 | 151 | 153 |
| 120 | 120 | 122 | 124 | 126 | 128 | 130 | 132 | 134 | 136 | 138 | 140 | 142 | 144 | 146 | 148 | 150 | 152 | 154 | 156 |
| 121 | 122 | 124 | 126 | 128 | 130 | 132 | 134 | 136 | 138 | 140 | 142 | 144 | 146 | 148 | 150 | 152 | 154 | 156 | 159 |
| 122 | 124 | 126 | 128 | 130 | 132 | 134 | 136 | 138 | 140 | 143 | 145 | 147 | 149 | 151 | 153 | 155 | 157 | 159 | 161 |
| 123 | 126 | 128 | 130 | 132 | 134 | 136 | 139 | 141 | 143 | 145 | 147 | 149 | 151 | 153 | 155 | 157 | 160 | 162 | 164 |
| 124 | 128 | 130 | 132 | 134 | 137 | 139 | 141 | 143 | 145 | 147 | 149 | 152 | 154 | 156 | 158 | 160 | 162 | 164 | 166 |
| 125 | 130 | 132 | 134 | 137 | 139 | 141 | 143 | 145 | 147 | 150 | 152 | 154 | 156 | 158 | 160 | 163 | 165 | 167 | 169 |
| 126 | 132 | 134 | 137 | 139 | 141 | 143 | 145 | 148 | 150 | 152 | 154 | 156 | 159 | 161 | 163 | 165 | 167 | 170 | 172 |
| 127 | 134 | 137 | 139 | 141 | 143 | 146 | 148 | 150 | 152 | 154 | 157 | 159 | 161 | 163 | 166 | 168 | 170 | 172 | 175 |
| 128 | 136 | 139 | 141 | 143 | 146 | 148 | 150 | 152 | 155 | 157 | 159 | 161 | 164 | 166 | 168 | 171 | 173 | 175 | 177 |
| 129 | 139 | 141 | 143 | 146 | 148 | 150 | 152 | 155 | 157 | 159 | 162 | 164 | 166 | 169 | 171 | 173 | 176 | 178 | 180 |

## CRITERIA FOR DIAGNOSIS

1 NORMAL HEART—The outline of the shadow of the heart can be divided into portions or curves each due to individual heart chambers or to great vessels In the P A view (Figs 1 7 8) on the right side the curve of the right atrium is immediately above the diaphragm and above this the curve formed by the ascending aorta An indentation frequently separates the two On deep inspiration a small triangular shadow sometimes may be seen between the diaphragm and the lower contour of the right atrium It is due to the inferior vena cava and is not to be mistaken for pleuro-pericardial adhesions Above the arch the vertical shadow of the superior vena cava often is noted to extend parallel and close to the spine

Three curves are noted on the left side Above is the convex knob of the aortic arch as it turns backwards and to the left Immediately below this is the outline of the pulmonary artery normally straight concave or slightly convex and often called the pulmonary artery segment Below this the lateral contour of the left ventricle extends to the diaphragm Occasionally the left auricle may occupy a position between the pulmonary artery and the left ventricle on this border When normal it rarely can be recognized except by electrokymography

The direction of the systolic pulsations of the left border of the cardiac shadow is inward over the left ventricle and outward and upward over the pulmonary artery segment A *point of adjacent opposite pulsations* can thus be identified The lower border of the heart even when viewed in deep inspiration cannot ordinarily be seen below the dome of the left side of the diaphragm In the cardio diaphragmatic angle a triangular shadow less dense than left ventricle and diaphragm is frequently seen and is due to epicardial fat (Fig 1)

In the right anterior oblique position (Figs 2 5) immediately above the diaphragm the anterior border of the cardiac shadow is formed by the anterior portion of the left or right ventricle depending upon the degree of rotation Above this is the shadow of the pulmonary artery and—highest—that of the ascending aorta The latter is continuous above with a foreshortened view of the transverse portion of the aortic arch The descending limb of the aorta often may be noted between the posterior surface of the heart and the spinal column It is best seen during inspiration and best in subjects with kyphosis or emphysema The posterior contour of the heart is formed below by the inferior vena

## TABLE II PREDICTION TABLE FOR AVERAGE NORMAL ORTHODIAGRAPHIC MEASUREMENTS OF THE TRANSVERSE DIAMETER OF THE HEART*

Predicted T D = + 0 1094 × A − 0 1941 × H + 0 8179 × W + 91 86 [illegible]

| I | | | | II | | | |
|---|---|---|---|---|---|---|---|
| Stature | | | Trans Diameter | Weight | | | Trans Diameter |
| Cm | In | Area Sq Cm | Mm | Kg | Pounds | Area Sq Cm | Mm |
| 150 | 59 | 66 7 | 66 71 | 50 | 110 | 17 00 | 40 90 |
| 151 | | 67 57 | 66 5 | 51 | 112 2 | 17 34 | 41 71 |
| 152 | 60 | 68 44 | 66 36 | 52 | 114 4 | 17 68 | 42 53 |
| 153 | | 69 31 | 66 16 | 53 | 116 6 | 18 02 | 43 35 |
| 154 | | 70 18 | 65 97 | 54 | 118 8 | 18 36 | 44 17 |
| 155 | 61 | 71 05 | 65 77 | 55 | 121 | 18 70 | 44 98 |
| 156 | | 71 92 | 65 58 | 56 | 123 2 | 19 04 | 4[illegible] 80 |
| 157 | | 72 79 | 65 39 | 57 | 125 4 | 19 38 | 46 62 |
| 158 | 62 | 73 66 | 65 19 | 58 | 127 6 | 19 72 | 47 44 |
| 159 | | 74 53 | 65 00 | 59 | 129 8 | 20 06 | 48 26 |
| 160 | 63 | 75 40 | 64 80 | 60 | 132 | 20 40 | 49 07 |
| 161 | | 76 27 | 64 61 | 61 | 134 2 | 20 74 | 49 89 |
| 162 | | 77 14 | 64 42 | 62 | 136 4 | 21 08 | 50 71 |
| 163 | 64 | 78 01 | 64 22 | 63 | 138 6 | 21 42 | 51 53 |
| 164 | | 78 88 | 64 03 | 64 | 140 8 | 21 76 | 52 35 |
| 165 | 65 | 79 7 | 63 83 | 65 | 143 | 22 10 | 53 16 |
| 166 | | 80 62 | 63 64 | 66 | 145 2 | 22 44 | 53 98 |
| 167 | | 81 49 | 63 45 | 67 | 147 4 | 22 78 | 54 80 |
| 168 | 66 | 82 36 | 63 25 | 68 | 149 6 | 23 12 | 5[illegible] 62 |
| 169 | | 83 23 | 63 06 | 69 | 151 8 | 23 46 | 56 44 |
| 170 | 67 | 84 10 | 62 86 | 70 | 154 | 23 80 | 57 25 |
| 171 | | 84 97 | 62 67 | 71 | 156 2 | 24 14 | 58 07 |
| 172 | | 85 84 | 62 47 | 72 | 158 4 | 24 48 | 58 89 |
| 173 | 68 | 86 71 | 62 28 | 73 | 160 6 | 24 82 | 59 71 |
| 174 | | 87 58 | 62 09 | 74 | 162 8 | 25 16 | 60 52 |
| 175 | 69 | 88 45 | 61 89 | 75 | 16[illegible] | 25 50 | 61 34 |
| 176 | | 89 32 | 61 70 | 76 | 167 2 | 25 84 | 62 16 |
| 177 | | 90 19 | 61 50 | 77 | 169 4 | 26 18 | 62 98 |
| 178 | 70 | 91 06 | 61 31 | 78 | 171 6 | 26 [illegible]2 | 63 80 |
| 179 | | 91 93 | 61 12 | 79 | 173 8 | 2[illegible] 86 | 64 61 |
| 180 | 71 | 92 80 | 60 92 | 80 | 176 | 27 20 | 65 43 |
| 181 | | 93 67 | 60 73 | 81 | 178 2 | 27 54 | [illegible] |
| 182 | | 94 54 | 60 53 | 82 | 180 4 | 27 88 | 67 07 |
| 183 | 72 | 9[illegible] 41 | 60 34 | 83 | 18[illegible] 6 | 28 [illegible] | 67 89 |
| 184 | | 96 28 | 60 15 | 84 | 184 8 | 28 [illegible]6 | 68 70 |
| 185 | 73 | 97 15 | 59 95 | 85 | 187 | 28 90 | 69 [illegible]2 |
| 186 | | 98 02 | 59 76 | 86 | 189 2 | 29 24 | 70 34 |
| 187 | | 98 89 | 59 56 | 87 | 191 4 | 29 58 | 71 16 |
| 188 | 74 | 99 76 | 59 37 | 88 | 193 6 | 29 92 | 71 98 |
| 189 | | 100 63 | 59 18 | 89 | 195 8 | 30 26 | 72 79 |
| 190 | | 101 50 | 58 98 | 90 | 198 | 30 60 | 73 61 |
| 191 | 75 | 102 37 | 58 79 | 91 | 200 2 | 30 94 | 74 43 |
| 192 | | 103 24 | 58 59 | 92 | 202 4 | 31 28 | [illegible] |
| 193 | 76 | 104 11 | 58 40 | 93 | 204 6 | 31 62 | 76 06 |
| 194 | | 104 98 | 58 21 | 94 | 206 8 | 31 96 | 76 88 |
| 195 | | 105 85 | 58 01 | 95 | 209 | 32 30 | 77 70 |
| 196 | 77 | 106 72 | 57 82 | 96 | 211 2 | 32 64 | 78 [illegible] |
| 197 | | 107 59 | 57 62 | 97 | 213 4 | 32 98 | 79 34 |
| 198 | 78 | 108 46 | 57 43 | 98 | 215 6 | 33 32 | 80 15 |
| 199 | | 109 39 | 57 23 | 99 | 217 8 | 33 66 | 80 97 |
| 200 | 79 | 110 20 | 57 04 | 100 | 220 | 34 00 | 81 79 |

To find normal transverse diameter for a given individual add T D figure for stature to T-D figure for weight and to the total add 1 mm for every decade of age e g height 6 feet weight 187 pounds age 50 = 134 8 mm T-D or 60 31 + 69 52 + 5

The figures of this table are valid for orthodiagrams and for male subjects The hearts of female subjects of same stature weight and age are slightly smaller in size Brunton has suggested to subtract 8 cm for T D of female

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zontally on an elevated diaphragm the anatomic apex of the heart is displaced to the left and the lower portion of the left contour is often obscured by the diaphragm (Fig 8) Relatively more of the contour of the left ventricle and less of the pulmonary artery segment can be seen

b *Vertical heart*—In *asthenic individuals* the position of the heart is vertical it appears long and narrow (Fig 7) In extreme forms it appears suspended it is drop or pear shaped (hypoplastic) The pulmonary artery forms the larger part of the left *contour below the aortic* knob In the right anterior oblique position the pulmonary artery is prominent so that in asthenic individuals this appearance does not *necessarily* indicate enlargement of the right ventricular outflow tract

c *Oblique or globular heart*—Between the extremes of horizontal and vertical shaped hearts is the oblique shape seen in sthenic individuals The relative length of the pulmonary artery contour and that of the left ventricle may be more or less *equal*

In infants the shape of the heart is usually globular It is centrally located and its borders reach about equally far to the left and right

2 Left Ventricle—The left ventricle anatomically and functionally can be subdivided into two portions the inflow and outflow tracts

a The *outflow tract* extends from the apex to the aortic valve and is the first portion of the left ventricle to become enlarged This is *manifested* roentgenologically by elongation of the left ventricular segment below the *point* of opposite pulsations Elongation may be recognized within the left diaphragmatic density or within accumulations of gas in the stomach or colon (Fig 9) or by an increase in the convexity of the upper portion of the left ventricle immediately below the point of opposite pulsations (Fig 10) Differentiation from a horizontally placed *normal* heart is accomplished by observing the effect of deep inspiration In a *normal* heart the unusual convexity should disappear

b The *inflow tract* extends from the mitral valve to the apex of the left ventricle Its enlargement occurs later than that of the outflow tract Roentgenologically the enlargement of the inflow tract is recognized by an increase in length and depth of the left ventricular segment seen in the left anterior oblique position (Fig 11) It changes from a shallow curve to a longer and more convex segment and bulges posteriorly The interventricular groove is displaced downward into the density of the infradiaphragmatic structures and cannot be identified

With marked enlargement when viewed in the P A position the left ventricular border is displaced towards the left lateral chest wall The rounding occupies the lower two-thirds or more of the left contour and

cava and a portion of the right atrium above this by the left atrium

The air column of the trachea and the right bronchus can be seen between the shadow of the descending portion of the arch of the aorta and the upper posterior outline of the heart The esophagus lies between the anterior surface of the descending aorta and the posterior surface of the heart When filled with a thick barium paste its course is seen to be essentially vertical forming a shallow curve posteriorly The continuity of its anterior contour is broken above by the normal indentations of the transverse part of the aorta below this by the right bronchus and finally by the constriction of the diaphragm The filling of the esophagus with barium paste should be a part of the routine examination by roentgenoscopy

In the left anterior oblique position (Figs 3 6) the anterior outline is formed from below upward by the right ventricle the right atrium and its auricle and the ascending aorta The contour of the right ventricle and the right atrium is vertical the right auricle may slope obliquely towards the aorta The ascending limb of the aortic arch is seen to curve upwards and posteriorly and is continuous with the transverse portion of the arch This curves downward into the descending limb of the arch and the thoracic aorta which partly overlap the shadow of the spinal column The posterior outline of the heart is formed by the left ventricle below and the left atrium above A shallow indentation the atrioventricular groove may separate the two The pulsations of the left ventricle are of greater amplitude At or near the junction of the left ventricle with the diaphragm an indentation may be observed which is plainest in systole during deep inspiration This is due to the interventricular septum This has a movement of its own whose direction is independent of the chambers to either side of it

The bifurcation of the trachea is seen within and below the density of the aortic arch The right bronchus appears foreshortened while the course of the left bronchus is more vertical than horizontal and forms an angle of less than 45 degrees with the line of the trachea depending upon the degree of rotation The left branch of the pulmonary artery occasionally may be seen as a shadow less dense and smaller than the aorta arching over the left bronchus and having its origin in a density within the heart shadow which represents the bifurcation of the trunk of the pulmonary artery The brachiocephalic vessel group mounted on top of the aortic arch forms the shadow anterior to the trachea

The appearance of the cardiac shadow is greatly influenced by the position occupied by the diaphragm

a *Horizontal heart* —In *hypersthenic individuals* the heart lies hori

viewed in the right anterior oblique position there may be an anterior bulge at a lower level than the pulmonary artery

b *Rotation of the heart on its vertical axis*—This occurs when the work of the right ventricle is increased out of proportion to that of the left ventricle Anterior structures move towards the left to form the left lateral contour while posterior structures such as the left atrium appear in the right contour and may be seen within the right portion of the heart shadow as an added or double density The pulmonary artery segment increases in size the prominence of the aortic knob diminishes the left ventricular contour becomes shorter and straighter (Figs 19A 20A)

c *Enlargement of the inflow tract*—The inflow tract is that portion of the chamber extending from the tricuspid valve to the apex Its enlargement produces increased depth of the chamber and is best seen in the left anterior oblique position In this position the diaphragmatic portion of the heart chiefly the right ventricle increases in length displacing the interventricular groove posteriorly and upwards During deep inspiration the diaphragmatic border remains elongated instead of diminishing as it does in the normal horizontal heart The bulge of the anterior contour toward the anterior chest wall is increased Marked enlargement of the outflow and inflow portions may occur without enlargement to the right in the P A view (Figs 16 17 18)

4 Left Atrium—Normally in the P A position the body of the left atrium does not appear The tip of the left auricle approaches and to a slight extent helps form the left contour between the pulmonary artery and left ventricle but rarely projects to form a definite contour of its own

a *Enlargement of the left atrium* is first and best recognized by its encroachment upon the retrocardiac space This is visualized in the right anterior oblique position as an arched density projecting towards the spine situated in the midportion of the posterior cardiac contour just below the tracheal bifurcation The extent of this horizontal enlargement can be inferred by the compression or displacement of the barium filled esophagus (Figs 19B 20P) One must be sure however to distinguish such displacement from that due to the esophagus being pulled to the left and backwards by adhesions to an elongated aortic arch or to other mediastinal structures (Fig 21) Since such an esophagus is pulled away from its usually intimate relationship to the posterior surface of the heart it can no longer be used as a guide to left atrial enlargement

b Superiorly the left atrium lies in relation to the main bronchi

may extend below the dome of the diaphragm In the left anterior oblique position the left ventricular contour extends more *posteriorly* (Fig 11) so that with the usual rotation it will markedly overlap the shadow of the spine The diagnosis of left ventricular enlargement should be made long before this stage is reached

c *Concentric hypertrophy* —When viewed in the P A position this will appear as a rounding of the left ventricular contour without elongation of this segment

d *Localized ventricular bulge* (ventricular aneurysm) —An abnormal bulging of the contour of the left ventricle due to thinning of a portion of the wall The most frequent site is at the apex and if slight *it is best seen on deep inspiration Other common sites are high* on the lateral wall (Fig 15) and on the posterior wall Obviously examination in the oblique as well as anterior positions is necessary

At the site of the aneurysmal bulge *there may be pericardial adhe*sions or an increase in density due to thrombus formation Rarely calcification is present Thinning of the entire wall of the left ventricle occasionally occurs and is difficult to differentiate from massive left ventricular enlargement except by electrokymography Aneurysmal bulges occur rarely in the right ventricle and the atria and diagnosis of such lesions is possible only when these areas are on the border of the cardiac silhouette

e *Localized impairment of contraction* is manifested either by a localized diminution of the amplitude of pulsations or by an actual reversal in the direction of pulsations (*paradoxical pulsations*) (*Figs* 12 13) This may indicate an area of acute myocardial disease or actual thinning of the wall due to scar formation

3 Right Ventricle —The right ventricle can also be divided into an inflow tract and an outflow tract

a *Enlargement of the outflow tract* —The earliest right ventricular enlargement occurs in the outflow tract This extends from the apex of the right ventricle to the pulmonic valve The first noticeable change when viewed in the P A position is a straightening or an increased prominence of the pulmonary artery contour *In the right anterior* oblique position there is seen a bulge of the pulmonary artery into the retrosternal space made even more prominent by elongation of the *conus portion of the right ventricle*

In certain forms of congenital heart disease the outflow tract may enlarge in the absence of pulmonary artery dilatation Under such conditions recognition of outflow tract enlargement is difficult and may require special techniques such as angiocardiography Sometimes when

erate size right atrium may be displaced to the right by a greatly enlarged right ventricle

6 SYMMETRICAL CARDIAC ENLARGEMENT —In the majority of cases of organic heart disease more than one chamber is found enlarged This is termed multiple chamber enlargement Enlargement of all the chambers is termed generalized cardiac enlargement When the enlargement of all the chambers is more or less symmetrical a uniformly acting systemic agent is usually found to be the cause for example anemia myxedema *et cetera*

One should not rely on characteristic configurations Terms like mitralization duck shape or *sabot* shape heart are undesirable for they do not give the information obtained from systematic description of the various chambers

7 DISPLACEMENT OF HEART —Slight displacement toward one side may simulate enlargement in this direction especially if associated with a scoliosis of the spine in the opposite direction Marked displacement of the heart may occur with pneumothorax pleural effusions atelectasis and pulmonary fibrosis Downward displacement at times to one side may occur with large aortic aneurysms or with an enlarged substernal thyroid gland The contours of the heart often cannot be distinguished from the shadow of dense surrounding structures Roentgenoscopic examination is necessary particularly in the oblique positions with an attempt to visualize the contours of the individual chambers and to rule out chamber enlargement. Angiocardiography is particularly valuable in these conditions

Deformities of the thoracic spine and of the chest wall not only displace but also distort the heart and aorta producing apparent enlargement of the heart or of individual chambers True enlargement may occur in certain cases

Scoliosis of the spine if convex toward the right displaces the heart into the left chest rotating it so that the anterior surface tends to appear on the left lateral contour and the posterior surface tends to form the contour on the right Scoliosis of the spine if convex toward the left displaces the heart into the right chest causing a rotation in the opposite direction Elevation of a portion of the diaphragm by gas eventration or abdominal fluid or herniation through the diaphragm also may displace and distort heart contours

Funnel-chest deformity if severe may compress the heart from the front flattening it by diminishing the antero-posterior diameter usually

below the bifurcation of the trachea *Vertical or upward enlargement* of the left atrium can be observed in its successive stages in the left anterior oblique position by a disappearance of the free space between the two main bronchi and a spreading of the angle between them Finally there is compression or displacement of the left bronchus upward and to the left (Figs 20C 22C)

c *Enlargement toward the right*—Enlargement of the left atrium may extend far enough to the right to appear in the middle portion of the right contour of the heart in the P A position (Figs 19A 20A) There may then be two more or less distinct curves the lower being the right atrium and the upper the left atrium Not infrequently the density of the body of the left atrium is visualized within the upper central portion of the heart The rotation of the heart associated with right ventricular enlargement accentuates this displacement of the left atrium towards the right

d *Enlargement toward the left*—Enlargement of the left auricle does not appear on the left cardiac border at an early stage since it is obscured by the coincident enlargement of the pulmonary artery Later a segment of a greatly enlarged left auricle may extend farther to the left than the pulmonary artery and form a recognizable contour of its own It may be identified by characteristic atrial wave like pulsations

5 Right Atrium—*Enlargement of the right atrium* occurs early in the trabeculate portion which lies anteriorly and lateral and quite late in the rest of the atrium

a *Enlargement of auricle*—Early enlargement is recognized by elongation of the right auricle whose contour is best seen in the left anterior oblique position (Figs 16D 21) Characteristic wave like pulsations may occasionally serve to identify this segment With marked right atrial enlargement this segment becomes less oblique and may even become horizontal (Fig 22C)

b *Enlargement posteriorly*—*The posterior smooth portion of the* atrium shows enlargement late It may be noted in the right anterior oblique position when a posteriorly convex shadow of increased density projects into the lower retrocardiac space immediately above the diaphragm replacing the former triangular density of the inferior vena cava Because of the position of the right atrium to the right of the esophagus the barium filled esophagus will be noted to cross through the shadow of the enlarged right atrium (Fig 22B) in contrast to the posterior displacement of the esophagus that occurs when left atrial enlargement occurs Enlargement to the right in the P A view is not a reliable criterion for enlargement of the right atrium Even a mod

in the presence of diminished pulsations in the rest of the cardiac contour should suggest the diagnosis. Differentiation from generalized cardiac enlargement is sometimes difficult. Effusions of less than 300 c.c. usually cannot be diagnosed.

b. *Adhesions* (Pericardial adhesions).—Adhesions between visceral and parietal layers are impossible to visualize. Adhesions between pericardium and pleura may cause a systolic retraction of visible pleura or lung tissue. Adhesions between the pericardium and diaphragm, ribs, sternum and vertebrae may be demonstrable with proper rotation of the patient. Pericardial adhesions may prevent the heart from shifting with changes in position but this feature may also be observed with unusually large hearts. An added linear shadow (Fig. 36) of increased density may occasionally be observed within the cardiac shadow adjacent to the border. This is due to a thickened pleuro-pericardial adhesion. Apparent elevation of the heart during deep inspiration may be observed when the pericardium is adherent to the mediastinal structures or vertebrae while the diaphragm descends with inspiration.

Pulsations at the site of adhesions may be diminished though the uninvolved portions of the heart may pulsate with normal or increased amplitude. Cardiac enlargement frequently occurs in association with pericardial adhesions to regional (extra-cardiac) structures.

c. *Constriction* (Constriction of the pericardium).—Dense adhesions may envelop the greater portion of the entire heart as a glove envelops the hand. These pericardial scars hamper diastolic expansion and may also affect systolic contraction. Over a period of time a diminution in cardiac size and the development of a more vertical position may occur. Diminished pulsations are difficult to substantiate because of the marked variability in the amplitude of cardiac pulsations from one heart to another. Roentgenkymography offers a graphic representation of diminution or of complete absence of pulsations over a portion of the heart and may also indicate a lessened period of diastolic relaxation and a plateau type of curve in the latter portions of diastole.

d. *Calcification*.—Calcification of the pericardium is recognized by dense linear, sometimes wave-like, shadows following the borders of the heart. It is frequently better seen in oblique positions (Figs. 37, 38, 39). Calcification of the pericardium may partly or completely encircle the base of the heart in the region of the atrio-ventricular groove and extend over the upper part of the right ventricle. In some cases enlargement of the right and left atria results (Fig. 39). The clinical picture of pericardial constriction may ensue though the heart is evidently enlarged.

e. *Cysts*.—Localized protrusions of the cardiac silhouette may be

displacing and rotating the heart into the left chest similar to the appearance with scoliosis of the spine convex to the right

Emphysema may cause enlargement of the outflow tract of the right ventricle which combined with a vertical position of the heart may greatly increase the pulmonary artery prominence Elevation of the heart and mediastinum from its diaphragmatic attachment due to bilateral upper lobe pulmonary fibrosis is occasionally seen

8 Calcification Intracardiac —Calcification of (a) the *heart valves* or (b) the *annuli fibrosi* may be noted on roentgenography or roentgenoscopy With good accommodation roentgenoscopy is the more reliable method Dense intracardiac linear or circular shadows may be seen moving up and down or in rotary fashion synchronous with cardiac pulsations This is best noted in oblique positions

c Calcification of the *coronary arteries* is rarely visualized A double row of linear densities with motion synchronous with cardiac pulsations may be visualized in regions where the main coronary arteries may be expected The most common site to find such calcifications is below and anterior to the ascending aorta in the right anterior oblique position

d Calcification of the *myocardium* may occur at sites of extensive myocardial necrosis such as for example with localized ventricular aneurysmal bulge

e *Endocardial* calcification is rare and represents the secondary *deposition of calcium on damaged endocardium* almost always on the posterior wall of the left atrium This is best recognized in the right anterior oblique position

9 Pericardium —a *Effusion* —*Pericardial effusion causes a generalized* enlargement of the cardiac area and a disappearance of the usual curves of the outline in the P A position (Fig 35) Diminution of pulsations is variable depending upon the amount of the effusion Filling the stomach with air may reveal absent or diminished pulsations of the inferior contour of the heart An increase in the acuteness of the right cardio diaphragmatic angle is common In the right anterior oblique position filling of the right posterior inferior recess in the vicinity of the inferior vena cava may be observed early in the course of the effusion The superior vena cava is usually dilated

Disproportionate widening of the upper contours in the P A view may be accentuated in the reclining position Occasionally decrease of the density at the base of the heart may be observed in the upright position as compared with that in the supine Normal aortic pulsations

indentation (Figs 24A B C D) This pulling is due to the normal adhesions between the esophagus and the elongated aorta at this level

Descending limb of aortic arch and thoracic aorta —Participation of this portion in generalized elongation of the aortic arch results in tortuosity In the left anterior oblique position a sinuous curvature or an apparent buckling may appear Dilatation of the descending aorta is frequently seen in elderly individuals The density of the shadow of the descending aorta in adults makes it readily visualized in the right oblique position It appears in the retrocardiac space distinguishable from the greater densities of the heart and the spinal column especially during deep inspiration In aortic insufficiency the descending aorta is rendered more visible even in children

Elongation and dilatation of the descending aorta results in a prominent curve with a convexity to the left seen in the P A view lying below the aortic knob and frequently extending beyond the hilus Below this density can often be followed within the cardiac shadow Frequently the entire aorta is diffusely dilated The aortic shadow then has a fusiform or spindle shaped contour

b Calcification of the aorta most frequently occurs in the descending and transverse portions In the P A position the superimposition of calcified plaques seen tangentially in the transverse portion are more readily noted in the aortic knob Calcification confined to the ascending aorta should be considered as almost diagnostic of syphilitic aortitis

c *Hypoplasia* —Hypoplasia of the aorta may occur as a solitary congenital anomaly Relative hypoplasia is not infrequently seen in mitral stenosis and in congenital anomalies when the volume flow of blood within the aorta is diminished The diagnosis may be made on fluoroscopy when the ascending and descending limbs of the aortic arch are unusually close together It may be confirmed by angiocardiography

II ANEURYSM OF AORTA —a *Saccular* Only a localized ballooning or bulge should be called an aneurysm (Fig 25) The diagnosis of aneurysm is justified only when the mass is seen to be a part of the shadow of the aorta when viewed from all directions The mass may show expansile or transmitted pulsations The absence or presence of pulsations however cannot be used as a final diagnostic criterion Calcification within the wall of the aneurysm is often present

Aneurysm of the ascending and transverse portions of the aorta may displace and constrict the trachea and bronchi and erode adjacent bony structures Huge aneurysmal sacs may entirely obscure large portions of the thorax A positive differentiation from pleural effusion or intrathoracic neoplasm may be difficult Angiocardiography may be expected

due to an encapsulated pericardial effusion to herniation or diverticulum formation or to fluid accumulation within an actual cyst It usually is ovoid or semicircular in shape is not opacified on angiocardiography and its pulsations are transmitted from the contiguous portion of the heart The actual etiological diagnosis usually is made at operation

10 Aorta —a —*Elongation and dilatation* —Elongation of the aorta (Figs 23 24) occurs in association with an increase of the arterial pressure or with an increased blood flow through this structure Under *such circumstances the vessel becomes too long between its fixation at* the base of the heart and the diaphragm There is usually an associated dilatation due to stretching of the wall Elongation with dilatation also occurs in hypertension and as a result of loss of elasticity due to arteriosclerosis or to intrinsic disease of the aortic wall such as syphilitic aortitis Detailed visualization of the entire thoracic aorta and its brachiocephalic branches is best achieved by angiocardiography

Ascending portion of aortic arch —The immediate supraventricular portion is hidden by the overlying right auricle and cannot be visualized except by angiocardiography Elongation of the upper portion of the ascending aorta is best seen in the left anterior oblique and posteroanterior positions In the left anterior oblique position the vertical course of the ascending limb is changed to an anterior convexity In the P A position the transverse portion of the arch is elevated to or above the level of the clavicles The aortic knob protrudes far into the left lung field The lower portion of the trachea is pushed to the right by the horizontal portion of the arch

*It is difficult to ascertain dilatation of the ascending portion of the* arch apart from elongation Measurement in both oblique positions is deceptive because of the adjacent position of the superior vena cava *except when the density of the aorta is markedly increased or in the* rare instances when calcification is found within its walls

Transverse portion of aortic arch —Elongation and dilatation is best observed *in the left anterior oblique position The curve of the arch is* wider with increased separation of the ascending and descending limbs The density of the dilated transverse portion of the arch may be so marked as to render it visible even *within the increased illumination* of the trachea The aortic indentation upon the esophagus is deepened and the diameter of the lumen at this point can accurately be measured from the esophagus to the outer margin of the aortic knob This diameter however is clinically unimportant More important is the pulling of the esophagus posteriorly and to the left below the aortic

physema and if of extreme degree sclerosis of the pulmonary artery may be inferred. Calcified plaques may be present.

b. The chief components of the hilar shadows are the *secondary branches* of the *pulmonary artery*. In the P A view these are usually better visualized on the right. The air space of the right lower lobe bronchus emphasizes the cardiac density medially and the descending branch of the right pulmonary artery laterally. On the teleoroentgenogram the normal width of the shadow of the upper portion of the right descending branch varies from nine to fourteen millimeters. Dilatation of the secondary branches of the pulmonary artery is best noted in the P A view.

Expansile pulsations of small amplitude are normally observed in hilar branches of the pulmonary artery. A marked increase in pulsations accompanied by rhythmic increase and decrease in density (*hilar dance*) is seen when the pulmonary artery pulse pressure is increased as in pulmonary valvular insufficiency or incompetency.

c. *Narrowing* of the pulmonary artery and its branches may be demonstrated in pulmonic or infundibular stenosis (Fig. 28).

13. Pulmonary Fields.—Pulmonary manifestations are common in heart failure. Blood vessels, parenchyma and pleura may be involved.

a. *Hilar dilatation*.—Pulmonary hypertension is indicated by increased width of the secondary branches of the pulmonary artery. In the presence of diffuse stasis the shadows of the secondary branches of the pulmonary artery may increase or diminish in size with increase or decrease of cardiac insufficiency.

b. Chronic diffuse *pulmonary stasis* is manifested by increased density of the entire lung field with contrast especially poor at the base. This density is due to vascular engorgement, interstitial alveolar edema and desquamated alveolar epithelium. At times chronic stasis may be localized to one or more lobes or portions of such lobes simulating inflammatory involvement (Fig. 10). Acute diffuse stasis may be indicated by radiating densities extending from the hilar regions in a butterfly like pattern (Fig. 11a). Acute stasis may be localized to portions of one or more lobes (Fig. 12a). A diffuse stippled or miliary appearance throughout the lungs may be due to localized small areas of fibrosis and alveolar collapse. This occurs after chronic diffuse stasis disappears (Fig. 13).

c. *Pleural involvement* is manifested by effusion or thickening, either along the chest wall or in the interlobal fissures. Pleural effusions may be generalized or localized and are frequently seen in the interlobar spaces.

to differentiate between an aneurysm and an adjacent mediastinal mass. In rare instances opacification is hindered by a narrowed orifice or thrombi.

b. *Fusiform aneurysmal dilatation is impossible to differentiate from* marked dilatation of the aorta except by angiocardiography.

c. *Dissecting hematoma*—(Fig. 26) The hematoma resulting from the rupture of the intima may extend within the media into the great vessels of the neck, up and down the aortic arch, and may extend into the abdominal aorta. There is widening of the involved portions of the aorta and brachiocephalic vessels which is particularly impressive if earlier comparison films are available.

Angiocardiography may indicate localized or diffuse thickening of the aortic wall at the site of the dissection, or an irregular or abrupt narrowing of the aortic lumen. Less frequently, contrast substance may appear within an arterial false passageway.

12. Brachiocephalic Arteries, Widening of.—Elongation and dilatation of the right innominate artery is recognized by a triangular *shadow extending upward and outward from the upper right contour* of the ascending aorta (Fig. 12). Its prominence is enhanced by upward displacement of the elongated and dilated ascending aorta.

Widening of the left subclavian artery may be recognized by a vertical density *extending upwards above the region of the aortic knob* just to the left of the sternum, later arching to the left. The most common cause for such widening is coarctation of the aorta (Fig. 33A).

13. Superior Vena Cava, Dilatation of.—The superior vena cava may be visible normally in the P-A view, although the curve of the ascending aorta is the more prominent. With increase in venous pressure the superior vena cava may appear as a vertical shadow of variable width parallel to the sternum extending above or lateral to the ascending aorta (Fig. 22A).

14. Pulmonary Artery.—a. *Dilatation of the trunk* of the pulmonary artery is seen in the P-A view as a bulge with its convexity to the left, appearing below the aortic knob (Fig. 29A).

*Dilatation of the primary branches* (right and left pulmonary arteries).—The left pulmonary artery is seen in the left anterior oblique position as a shadow of increased width and density obliquely crossing the left bronchus (Fig. 29C). It emerges from a centrally placed density due to the bifurcation of the trunk of the pulmonary artery. *Markedly increased density* occurs most often in the presence of em

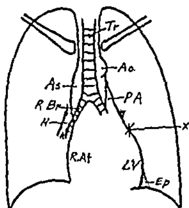



Fig 3 Normal Configuration (P A)
X—Point of opposite pulsations
Tr—Trachea
R Br—Right Bronchus
As—Ascending Aorta
H—Hilar (Secondary) branch of the Pulmonary Artery
Ao—Aortic Knob
P A—Pulmonary Artery
Ep—Epicardial fat

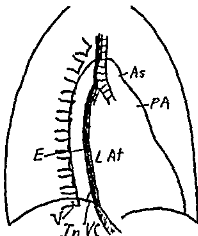



Fig Normal Configuration (R A O)
E—Esophagus
V—Vertebral column
In V C—Inferior Vena Cava
L At—Left Atrium
As—Ascending Aorta
P A—Pulmonary Artery

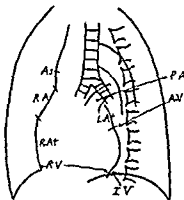



Fig 6 Normal Configuration (L A O)
A—Ascending Aorta
R A—Right Auricle
R At—Right Atrium
R V—Right Ventricle
P A—Pulmonary Artery
L At—Left Atrium
A V—Atrio Ventricular groove
I V—Interventricular groove

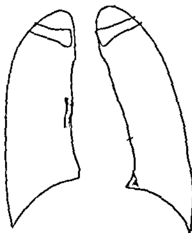


Fig 7 Vertical Heart

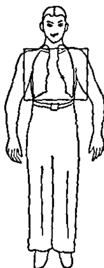


Fig. 1. Postero-Anterior (P. A.) Position. Fluoroscopic screen against the anterior chest wall.



Fig. 2. Right Anterior Oblique (R. A. O.) Position. Arms may be abducted or elevated and held on top of the head.



Fig. 3. Left Anterior Oblique (L. A. O.) Position.

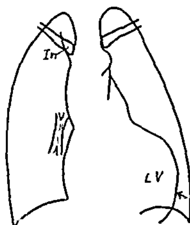



Fig 1_ Marked Enlargement of the Left Ventricle High left ventricular rounding The arrow points to a slight indentation best noted on fluoroscopy when the normal segment above this indentation moves inward during systole while the portion below the indentation moves outwards (paradoxical pulsation)

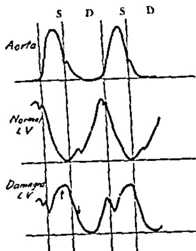



Fig 13 Electrokymogram In systole the aorta moves outward (up in the diagram) The normal portion of the left ventricle moves inward (down) In diastole (aortic notch indicating the closure of the aortic valve) the aortic motion is inward that of the normal left ventricle outward In the damaged portion of the left ventricle there is expansion in systole (outward motion first arrow) and inward recoil in diastole (second arrow) S Systole D Diastole

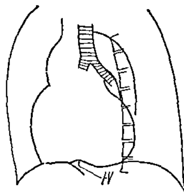



Fig 11 Marked Enlargement of the Inflow Tract of the Left Ventricle It overlaps the spine posteriorly and displaces the interventricular groove (IV) far anteriorly Note also the increased curvature (elongation) of the ascending aorta and the separation of the anterior and posterior limbs of the aortic arch indicating elongation of the transverse portion

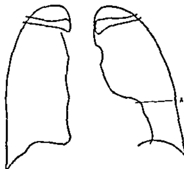



Fig 15 Ventricular Aneurysm This site of localized bulge is less frequent than is the lower portion of the left ventricular contour (pulsating outward in systole) as in Fig 1?

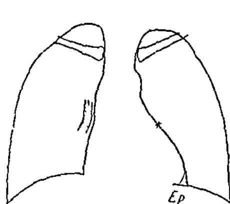



Fig. 8. Horizontal Heart.
X—Point of adjacent opposite pulsations.
Ep—Epicardial fat.

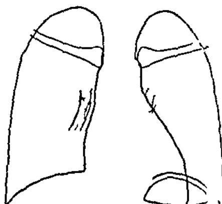


Fig. 9. Elongation of the Outflow Tract of the Left Ventricle. The length of segment below X is increased and extends below the dome of the left leaf of the diaphragm.

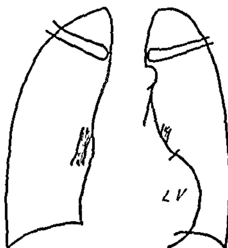



Fig. 10. Elongation of the Left Ventricle as expressed in rounding of the upper left ventricular contour and in the extension of the left ventricular density below the diaphragm.

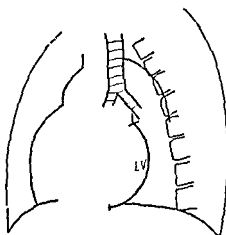
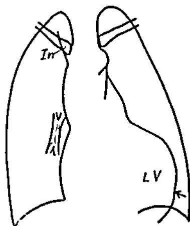
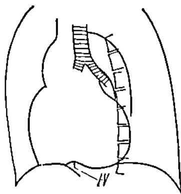


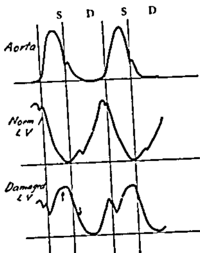

Fig. 11. Enlargement of the Inflow Tract of the Left Ventricle. Early elongation of this portion is shown by increased rounding in the LAO position. The interventricular groove is displaced downwards and anteriorly.



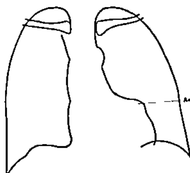

Fic 1 Marked Enlargen ent f the Left Ventricle High left ventricular rounding The arrow points to a slight indentation best noted on fluoroscopy when the normal segment above this indentation moves inward during systole while the portion below the indentation moves outwards (paradoxical pulsation)




Fi 11 Marked Enlargement of the Inflow Tract of the Left Ventricle It overlaps the spine posteriorly and displaces the interventricular groove (I V) far anteriorly Note also the increased curvature (elongation) of the ascending aorta and the separation of the anterior and posterior limbs of the aortic arch indicating elongation of the transverse portion




Fi 13 Electrokymogram In systole the aorta moves outward (up in the diagram) The normal portion of the left ventricle moves inward (down) In diastole (aortic notch indicating the closure of the aortic valve) the aortic motion is inward that of the normal left ventricle outward In the damaged portion of the left ventricle there is expansion in systole (outward motion first arrow) and inward recoil in diastole (second arrow) S Systole D Diastole




Fi 1 Ventricular Aneurysm This site of localized bulge is less frequent than is the lower portion of the left ventricular contour (pulsating outward in systole) as in Fig 12

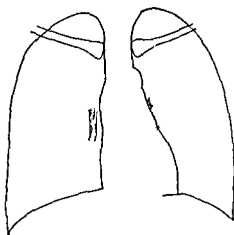


FIG 16A Vertical Heart (associated with bronchiectasis emphysema and pulmonary ventilation insufficiency) before enlargement of the right ventricle

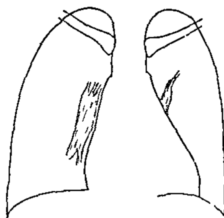


FIG 16B Enlargement of the Right Ventricle Same heart in right sided failure Note the disappearance of the aortic knob (rotation) plus straightening of the left upper contour Note also the increased width of the right hilar branch of the Pulmonary Artery

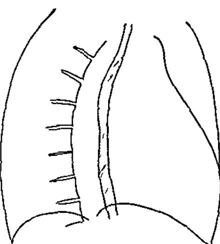


FIG 16C Right Anterior Oblique Position Note partial obliteration of the retrosternal space by the enlarged right ventricular outflow tract plus dilated pulmonary artery

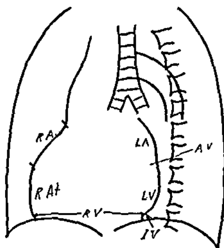



FIG 16D Enlargement of the Right Ventricular Inflow Tract and also of the Right Auricle (formerly termed the right auricular appendage) Increase in the length of the diaphragmatic portion indicates inflow tract enlargement

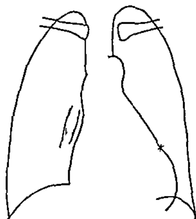
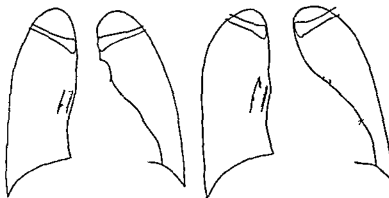


Fig 1 Right Ventricular Enlargement in a case of essential hypertension the pulmonary artery segment is prominent the left ventricle is moderately enlarged



Figs 18A and 18B Progressive stages in the Enlargement of the Right Ventricle leading to a disappearance of the aortic knob and rotation of the heart

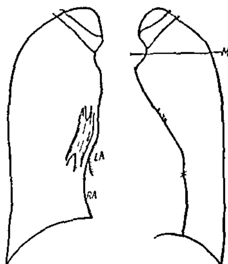



FIG 19A Enlargement of the Outflow Tract of the Right Ventricle Note the drop in convexity at X indicating that the left ventricle does not participate in the enlargement The increased density of the left atrium appears on the right contour
M—Manubrial Portion of the sternum

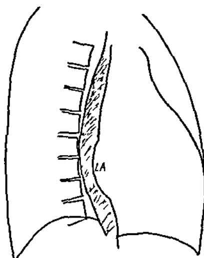



FIG 19B Same in the Right Anterior Oblique Position The pulmonary artery and the right ventricular outflow tract bulge into the retrosternal space The barium filled esophagus is indented by the enlarged left atrium

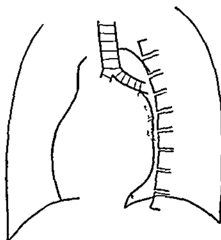


FIG 19C Same in the Left Anterior Oblique Position Only slight right ventricular outflow tract enlargement The left main bronchus is elevated by left atrial enlargement

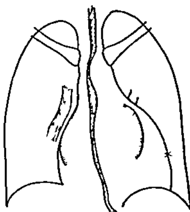


Fig 20A Enlargement of the Left Atrium appearing in the Postero Anterior View as an area of increased density forming the upper part of the right cardiac contour and indenting the barium filled esophagus slightly to the right The upper indentation in the esophagus is that of the aortic knob Note the small left ventricular contour

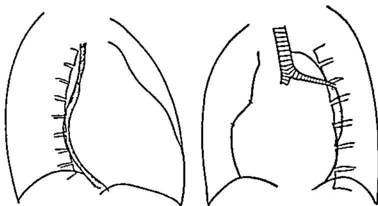


Fig 20B Same in the Right Anterior Oblique Position Marked left atrial enlargement also bulging into the retrosternal space due to right ventricular outflow tract enlargement plus pulmonary artery dilatation

Fig 20C Same in the Left Anterior Oblique Position The left main bronchus is elevated and compressed The left ventricular contour is not enlarged Thus cardiac enlargement in the P A view (Fig 20A) is due not to left ventricular enlargement but to displacement of this ventricle to the left by the enlarged right ventricle

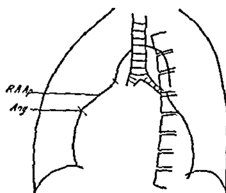



Fig 21 Enlargement of the Right Auricle (labeled R A App) L A O Position Elongation of the oblique *segment* lying between the ascending aorta and the lower anterior contour (right atrium above right ventricle *below*)

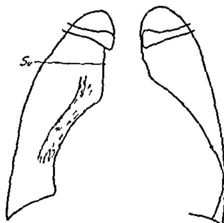



Fig 22A Right Atrial Enlargement A prominent right contour may be due to actual enlargement *of the right atrium to* displacement of the right atrium by right ventricular enlargement or to a combination of the two as in this instance
Su—Dilated Superior Vena Cava

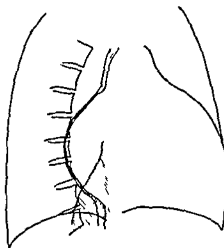


Fig 22B Enlargement of the Right Atrium posteriorly R A O Position This portion is shaded and seemingly is transected by the barium filled esophagus which actually is displaced pos*teriorly* by the only posterior heart chamber in contact with it the greatly enlarged left atrium

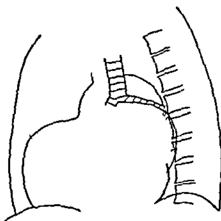


Fig 22C Marked Right Auricular (old terminology Right Auricular Appendix) Enlargement This segment is practically horizontal Note also the elevation and compression of the left main bronchus by the markedly enlarged left atrium

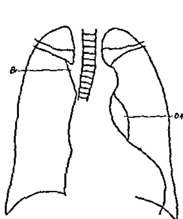



Fig 23A Elongation and Tortuosity of the Aorta and Brachiocephalic Vessels (Br) Note the increased curvature of the ascending aorta the prominent aortic knob and the tortuous descending portion of the arch and the thoracic aorta The lower portion of the trachea is pushed to the right by the elongation of the aorta

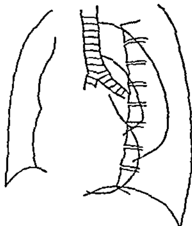


Fig 23B Same in the Left Anterior Oblique Position The elongation and tortuosity cause an apparent buckling or angulation of the aorta This is not real angulation since the course of the aorta turns at this point away from the observer

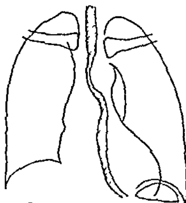


Fig 24A Displacement of the Barium Filled Esophagus to the Left Elongation of the transverse portion of the aortic arch pulls the adherent esophagus away from its usual relation to the left atrium When this is present the barium filled esophagus no longer may be used as a guide to left atrial enlargement

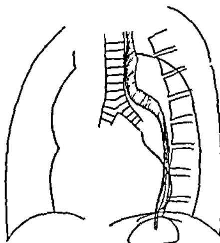


Fig. 21B Same in the Left Anterior Oblique Position. Posterior displacement of the barium filled esophagus. Note that the descending aorta is no longer prevertebral but is now paravertebral.

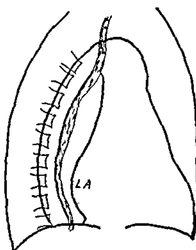



Fig. 21C Same in the Right Anterior Oblique Position. The barium filled esophagus is pulled posteriorly because of its aortic adhesions. This simulates posterior displacement by an enlarged left atrium which in this instance actually is enlarged but not sufficiently so to indent or displace the esophagus.

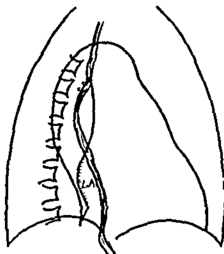
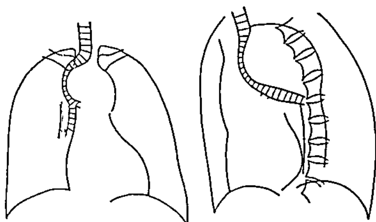



Fig. 21D A similar case in the Right Anterior Oblique Position. The esophagus is pulled posteriorly above, then pushed anteriorly below by the tortuous thoracic aorta. Nevertheless left atrial enlargement can be identified *but without the aid of the* barium filled esophagus.



Fi ..5A Aneurysm of tl Frai erse Lortion of tl e Aortic Ar h

Fi B Same in tl e Left An ter r Ol li que Losition

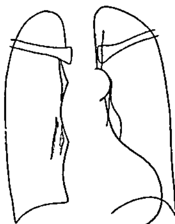


Fig 9C Dissecting Aneurysm of the Aorta The sha led por tions represent areas of d ssec tion within the walls of the aorta and the brach ocephalic vessels

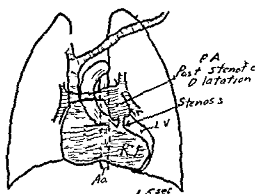



Fig 27A Angiocardiogram Tetralogy of Fallot (Infundibular stenosis with post-stenotic dilatation) The aorta is practically transposed receiving almost all of its blood from the right ventricle

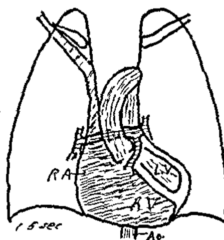



Fig 27B Angiocardiogram of the Heart in Tetralogy of Fallot (Infundibular plus valvular stenosis) The blunted appearance of the lowermost left contour is due to right ventricular enlargement which also displaces the interventricular septum and the left ventricle upwards and backwards Though the shunt of blood through the septal defect is from the right to the left ventricle opacification of the latter may be good fair or even poor

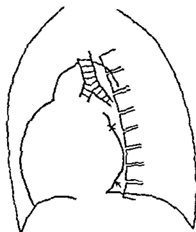


Fig 27C Same in the Left Anterior *Oblique Position* The increased size of the right ventricle and right atrium are seen *here The left ventricle* is displaced posteriorly and upward above the arrow which denotes the *interventricular groove*

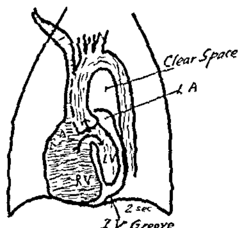
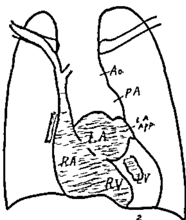



Fig 27D Angiocardiogram of same in the Left Anterior Oblique Position The pulmonary artery is not opacified thus the clear space below the aortic arch




Fi 98 An iocard ogram of Pulmonic (Valvular) Stenosis plus Atrial Septal Defect The pulm nary arter trunk is prominent bevon I the val ular steno s (post stenotic d lata tion) Note that this dilatation does not extend to the hilar *branches of the* pulm nary artery

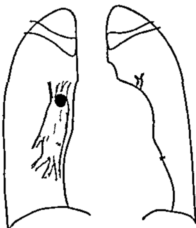
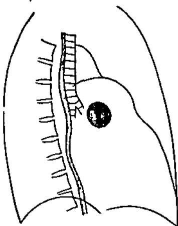


Fig 99A Atrial Septal Defect Note the tremendous dilatation of the secondary branches of the pulmonary artery The sha led area with n these vessels is due t an enlarged tertiary branch seen on en l The pulm nary ar tery tr nk is prominent tl e left ventricle is small



Fi 99B S me n the R t Ante Obl que Position The slight left atr al enlargement h s due to an assoc at l n t al stenos s (Lutembacher s Syn lrome) The haded a ea is due to the right branch of the pulmonary artery seen on end

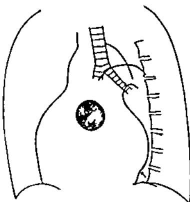


Fig 99C San e in the Left An ter or Oblique Posit on The shaded area is due to the right branch of the pulm nary a tery seen on en l Note that tl e p l monary artery is wider than the aorta The ht entricle an l right atrium are markedly en larged the left ventr cle is small

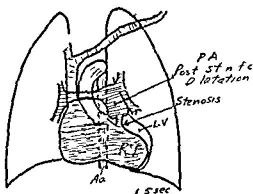



Fig 27A Angiocardiogram *Tetralogy of Fallot (Infundibular stenosis with post stenotic dilatation)* The aorta is practically transposed receiving almost all of its blood from the right ventricle

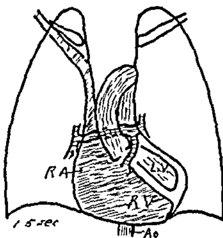



*Fig 27B Angiocardiogram of the Heart in Tetralogy of Fallot* (Infundibular plus valvular stenosis) The blunted appearance of the lowermost left contour is due to right ventricular enlargement which also displaces the interventricular septum and the *left ventricle upwards and backwards* Though the shunt of blood through the septal defect is from the right to the left ventricle opacification of the latter may be good fair or even poor

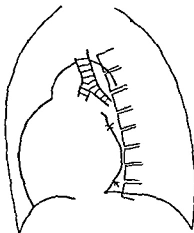


*Fig 27C* Same in the Left Anterior Oblique Position The increased size of the right ventricle and right atrium are seen here The left ventricle is displaced *posteriorly* and upward above the arrow which denotes the interventricular groove

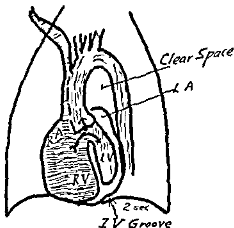



Fig 27D *Angiocardiogram of* same in the Left Anterior Oblique Position The pulmonary artery is not opacified thus the clear space below the aortic arch

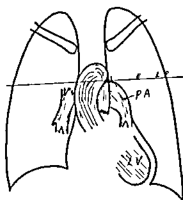



Fig 3 A Angiocardiogram in Patent Ductus Arteriosus Note the elevation of the left pulmonary artery (L P A) the re-opacification of the pulmonary arteries from the aorta

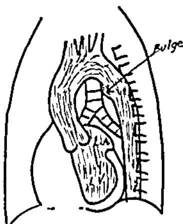



Fig 3-B Same in the Left Anterior Oblique Position The infundibular bulge on the anterior wall of the descending arch is characteristic though variants of this bulge may be confusing In this instance reopacification is not demonstrable

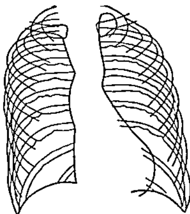


Fig 33A Coarctation of the Aorta The left ventricle is rounded and enlarged There is an absence of the aortic knob in the presence of an elongated ascending aorta and widened innominate and left subclavian arteries Erosion of the ribs is present

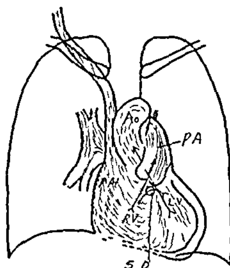



FIG 30 Angiocardiogram of Eisenmenger's Syndrome Arrows indicate the flow of blood from the left ventricle into the aorta and through the ventricular septal defect (SD) into the right ventricle The shunt here is from left to right thus the left ventricle and aorta are not opacified

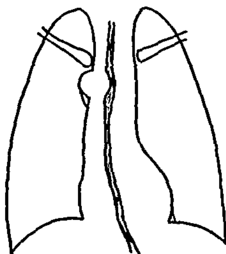


FIG 31A Right Aortic Arch (High crossing) Note the aortic indentation on the esophagus from the right

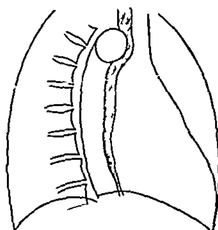


FIG 31B Same in the Right Anterior Oblique Position Note *the compression of the esophagus* from behind by the high aortic crossing to the left

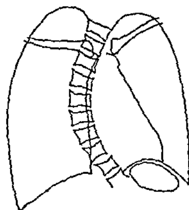


Fig 34 Cardiac Displacement (Scoliosis of the Spine) The heart is displaced into the left chest also rotated Frequently due to associated emphysema right ventricular outflow tract enlargement and pulmonary artery prominence may be demonstrated.

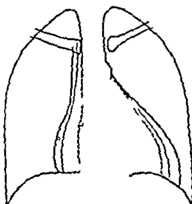


Fig 35 Pericardial Effusion Increase in size and regression occurring within two months Note the increased width just above the diaphragm the loss of individual chamber contours the increased acuity of the right cardio hepatic angle and dilatation of the superior vena cava

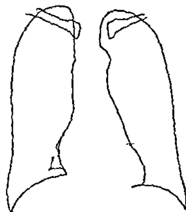


Fig 36 Pericardial Adhesions Irregularity in the cardiac contour whose motion is synchronous with the movement of the heart.

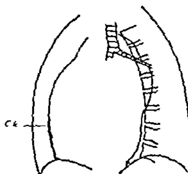



Fig 37 Localized Calcification of the Pericardium (L A O) This occurs in rheumatic as well as in non rheumatic hearts most frequently over the right ventricle

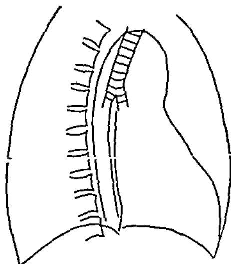


Fig 33B Same in the Right Anterior Oblique Position Note the elongated ascending aorta in contrast to a narrow descending aorta

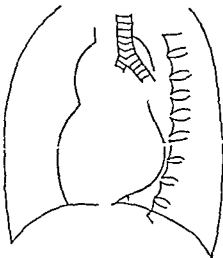


Fig 33C Contrast elongated and dilated Ascending Aorta with a narrow Descending Aorta

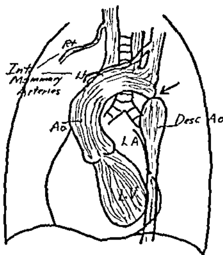
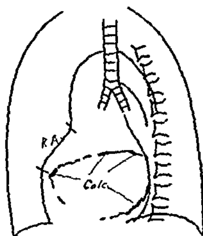



Fig 33D Angiocardiogram of same (L A O) The site of constriction post stenotic dilatation and some of the collaterals are demonstrated

39A PA position

39B R A O position




39C L A O position

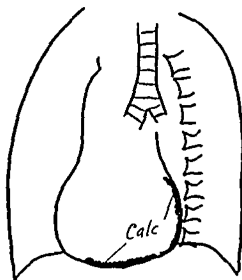
Fig. 39 Atrio-ventricular Ring Type of Pericardial Calcification. The calcification [illegible] in some contours but not in others. In the R A O position it frequently seems to be linear or L shaped. In the L A O position linear or punctuate calcific [illegible] within the heart shadow rather than at the contours. The heart [illegible] slightly or moderately enlarged chiefly the atria.

Calc

38A P A position

Calc

38B R A O position




38C L A O position

Fig 38 Diffuse Type of Pericardial Calcification Small heart, no demonstrable chamber enlargement, calcification of contours noted in all positions



FIG 41A Acute diffuse pulmonary stasis (pulmonary edema) in a 20 year-old male with *mitral stenosis* and insufficiency *Note* the butterfly like radiation extending from the hilar regions

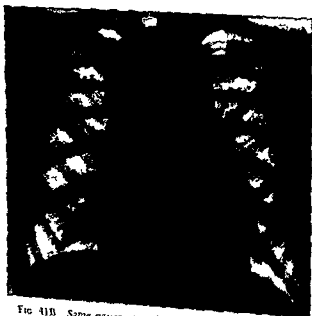


FIG 41B *Same patient two days later* The manifestations of acute diffuse pulmonary edema have disappeared



Fig 40 Chronic diffuse pulmonary stasis in a 58 year old male due to myocardial infarction and left ventricular failure In addition to diffuse increased density there is also a localized area of density in the right mid lung field



Fig 43 Diffuse stippled appearance due to small localized areas of alveolar collapse and fibrosis This miliary appearance is most common in the interval between attacks of acute or chronic diffuse pulmonary stasis



Fig. 12A. Acute localized pulmonary stasis in a 26 year old female with mitral stenosis. Homogeneous density is noted in the right lower lung field and a smaller discrete area on the left side.

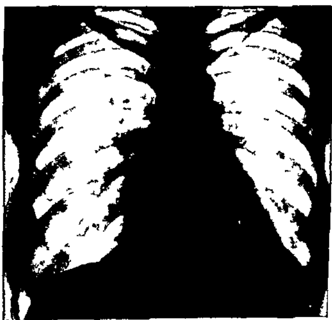
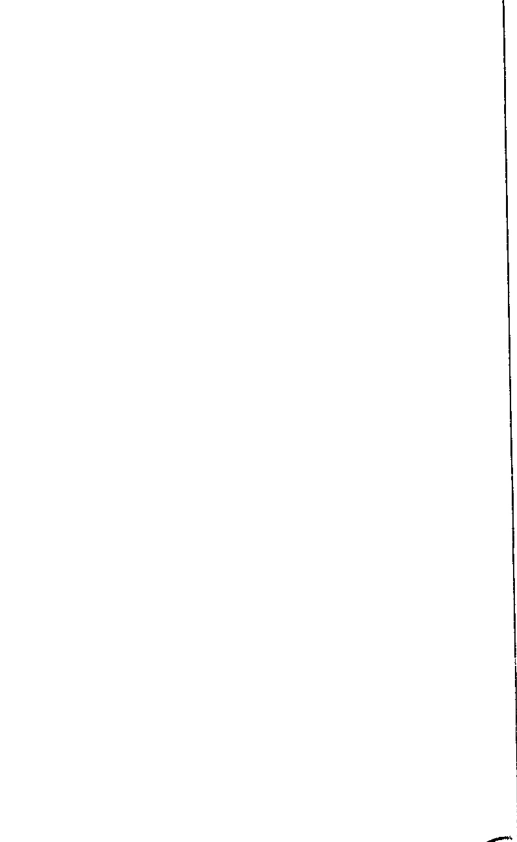


Fig. 12B. Same patient on the following day. The localized densities have disappeared.

# CRITERIA FOR ELECTROCARDIOGRAPHIC DIAGNOSIS



# CRITERIA FOR ELECTROCARDIOGRAPHIC DIAGNOSIS

# CRITERIA FOR ELECTROCARDIOGRAPHIC DIAGNOSIS

## CRITERIA FOR INSTRUMENTS*

### Electrocardiographs

1 Electrocardiographs shall be equipped with a suitable recording mechanism

2 The recorded response of the instrument to externally applied square wave voltages shall be adjustable to a sensitivity of 1 cm per millivolt when this voltage is applied to the leads of the instrument through a series resistance of 2 000 ohms This sensitivity shall be maintained without further adjustment within ±5 percent for a minimum period of three minutes under operating conditions Operating conditions for the purpose of this requirement are defined as follows (1) For instruments powered by alternating current at a specified power frequency the line voltage may vary from 105 to 130 volts and the power frequency ±2% from its specified value (2) For instruments powered by one or more batteries the voltage across the terminals of any or all when under operation should be between 80% and 100% of their rated voltage

Under these conditions the response of the instrument to its incorporated standardizing signal of 1 millivolt shall be within ±5 percent of the response to the externally applied test signal The instrument shall incorporate means of superimposing its intrinsic test signal upon the cardiographic tracing as recorded from any lead position It must be possible to apply this signal voltage continuously for a period of two or more seconds

3 When the instrument is standardized from a 1 cm response to 1 millivolt the deflection resulting from a sinusoidal voltage varying from 1 cycle to 15 cycles per second shall not fall below 90 percent and from 15 to 40 cycles per second shall not fall below 80 percent of the square wave response to equivalent voltage variation ** The amplitude response of the instrument to 1 millivolt peak sinusoidal voltage variation

* Modified from a report of the Advisory Committee on Electrocardiographs [illegible] Council on Physical Medicine and Rehabilitation of the American Medical Association [illegible] March [illegible]

** The Criteria Committee regards these as minimal frequency characteristics [illegible] have an amplitude response to a [illegible] sinusoidal voltage [illegible]

up to 300 cycles per second shall not exceed 100 percent of the square response to equivalent voltage variations.

4. The response of the instrument at 0.2 second after the application of a direct voltage of 1.0 millivolt shall not deviate more than ±10 percent from the response at 0.01 second. The test voltage of 1 millivolt should be applied to the leads of the instrument through a series resistance of 2,000 ohms.

5. When the instrument is adjusted to the sensitivity specified in requirement 2 the recorded response shall be directly proportional to the applied voltage (direct current) within ±5 percent over a range of 3 cm. on either side of zero.

6. With the two input terminals connected together, a potential difference applied between them and ground should not produce a deflection of more than 1 percent of that produced by the same potential difference applied between the two input terminals.

7. The instrument shall incorporate a means of continuously recording time intervals on the record and this must be by means of a device operating independently of the record driving mechanism. These intervals shall be of one second duration or less and the timing device shall be accurate within ±2 percent. However, a means of superimposing this time signal on the electrocardiographic tracing at the operator's will will be accepted as fulfilling this requirement. It should be possible to record the time signal for a period of at least two seconds. Recording paper preruled so as to indicate time intervals, assuming a constant paper speed, shall not be construed as fulfilling this requirement.

### VECTORCARDIOGRAPHS

Requirements for acceptable vectorcardiographs using cathode ray tubes have not yet been defined. Further investigation is pending.

# CRITERIA FOR TECHNIQUES

GENERAL

Changes in the electrocardiogram occur in variable degree with change in posture after meals after smoking after exercise and with the use of drugs The position of the body at the time the recording is made should be stated in the report The preferred position of the patient is recumbent The reclining table should be long enough and wide enough to support all extremities To eliminate shivering the temperature of the room in which the records are made should be comfortable For precise comparative work the patient should be in the basal state at each recording

BIPOLAR EXTREMITY LEADS

Bipolar extremity leads record the difference in potential between two extremities by connecting each to one of the input terminals of the recording device A lead such as this in which both electrodes are distant from the heart is described as bipolar The difference in potential between the Left arm and the Right arm is designated as Lead I between the Left Leg and the Right Arm as Lead II and between the Left Leg and the Left Arm as Lead III In each instance the connections to the galvanometer are to be made in such a way that positivity of the first named extremity with respect to the second results in an upright deflection in the finished record

In taking these leads the sensitivity of the recording instrument should be so adjusted that introduction of one millivolt in the circuit results in a deflection of one centimeter In practice this reference voltage or standardization should be recorded at the beginning and at the end of each lead

The electrodes may be placed on any part of the arms or of the left leg in making these leads so long as they are below the shoulders in the former and below the inguinal fold anteriorly and the gluteal fold posteriorly in the latter Any other placement of the electrodes made necessary by deformed or missing extremities must be noted on the record

The electrodes may be any of several types provided that a low resistance can be obtained between the surface of the electrode and the skin and so long as the metal is of low resistance and displays no condenser properties A convenient electrode to use on the extremities

*is one of German silver 3 5 cm × 5 0 cm* which is held in place by an elastic band and which makes contact with the skin through a conducting jelly

Electrodes and their contacts with the lead in wires of the recording device must be kept scrupulously clean The entire circuit from patient to machine must be inspected frequently for defects

UNIPOLAR EXTREMITY LEADS

The potential of any extremity may be obtained by connecting its electrode (exploring electrode) to one input terminal of the recording device The other terminal is connected to an indifferent electrode preferably one with a potential as close as possible to the mean potential of the body during the cardiac cycle Such an electrode can be constructed by connecting the right arm the left arm and the left leg to a central terminal each through a fixed non inductive resistance of 5 000 or more ohms It is imperative that the resistances between each extremity and the central terminal be equal A galvanometric lead in which the central terminal is used as the indifferent electrode is described is unipolar

Augmentation whereby the resulting deflections are one and one half times as large as their true size may be accomplished by severing the connection between the central terminal and the extremity being studied

In recording the extremity potentials either in the ordinary unipolar or augmented way the electrocardiograph is to be so adjusted that a deflection of one centimeter in the finished record corresponds to a potential difference of one millivolt Any increase in sensitivity of the instrument made necessary by small deflections should be clearly recorded on the curve preferably at the beginning and at the end of the lead Connections to the galvanometer are to be made in such a way that relative positivity of the exploring electrode will cause an upright deflection in the electrocardiogram

When made in the ordinary way the records from the right arm the left arm and the left leg are to be designated by the symbols $V_R$ $V_L$ and $V_F$ respectively *When the records have been augmented each* of these symbols should be preceded by a lower case letter a as follows $aV_R$ $aV_L$ $aV_F$

PRECORDIAL LEADS

The exploring or precordial electrode should be circular and 3 cm or less in diameter In children under ten years the diameter should be 1 5 cm or less It is desirable that multiple precordial leads be taken

and that the position of the precordial electrode be indicated by a subscript after the lead according to the following plan

Subscript 1 shall be used for a lead from the right sternal margin at the fourth intercostal space Subscript 2 for a lead from the left sternal margin at the fourth intercostal space Subscript 4 for a lead from the fifth intercostal space where it is crossed by the midclavicular line Subscript 3 for a lead from a point midway between points 2 and 4 Subscript 5 for a lead from the junction of the left anterior axillary line with the horizontal level of position 4 Subscript 6 7 and 8 are for leads on the same horizontal level but at the left midaxillary line (6) the left posterior axillary line (7) and the left midscapular line (8) respectively When additional leads are made from the right side of the thorax their location is to be indicated by arabic subscripts as for the left side to be followed by the letter R (for right) A lead from the fifth intercostal space in the right midclavicular line thus will bear the designation $V_{4R}$

If vertical deviations in the placing of the electrodes are made for any reasons the exact level should be indicated by the Roman numeral of the intercostal space following the Arabic numeral of the position e.g. $V_4$III would indicate the lead from the third intercostal space above the position of $V_4$

For routine purposes leads should be made from at least three areas widely distributed over the precordium In this regard the combination of locations 1 3 and 5 have been found useful though leads from all six positions are more informative and therefore preferable

*The indifferent or distant contact is to be placed on the central terminal* Leads *obtained* in this way are designated by the letter V followed by a *subscript* depending upon the *location* of the exploring electrode as described above

Leads made with the distant *contact* placed upon the left leg have been widely used They are designated by the letters CF followed by the numeral indicating chest position as above described Leads with the distant contact placed upon the right *arm* have been less used They are designated by the letters CR followed by the appropriate numeral The use of the central terminal is recommended because uniformly it will give a more nearly exact record of the precordial potentials than is obtained when using either of the limbs as the site for the distant contact *

* Editors Note Attention should be called to the fact that several articles in the literature have pointed out that at times when the myocardium is certainly the seat of a disease process the T wave may be inverted in leads CF and $CF_6$ but not in $V_5$ and $V_6$ This has been found in 6 of a series of 63 cases with diagnosed heart disease —Trans Assoc Amer Phys 19 1 61 381

In taking precordial leads connections to the recording device are to be made as described for the unipolar extremity leads so that relative positivity of the exploring electrode is represented by an upward deflection in the finished record

It will be found convenient in taking precordial leads with large deflections to adjust the electrocardiograph so that a deflection of one half centimeter in the finished record corresponds to a potential difference of one millivolt The sensitivity should be clearly indicated on the curve by recording at its beginning and at its end the effect of introducing a potential difference of one millivolt into the galvanometric circuit

### Esophageal Leads

The exploring or esophageal electrode is usually a small cylinder approximately 3 mm x 4 mm made of non corrosive metal It is connected by insulated wire of approximately 100 cm in length to a clip or jack to which an input terminal of the galvanometer may be attached readily Several ring electrodes insulated from each other at distances of 2 5 to 5 0 cm may be arranged along a small bore tube similar to a stomach tube With this electrode multiple leads may be made from several esophageal levels without moving the tube

The depth of an electrode in the esophagus is measured from the external nares In adults the exploring electrode is usually close to the heart at levels between 30 cm and 55 cm from the external nares Placement and localization are accomplished best with the aid of a fluoroscope

The central terminal is to be used as an indifferent electrode as described for other unipolar leads Response of the electrocardiograph to the introduction of a potential difference of one millivolt must be adjusted to the size of the deflections to be recorded and must be clearly shown on each lead made

The leads obtained are designated by the symbol V followed by the subscript E and an Arabic number to indicate the distance of the electrode from the external nares *e g* $V_{E}$[illegible] $V_{E}$[illegible]

### Vectorcardiograms

There is no agreement at present on the technique to be used in recording the vectorcardiogram in several planes However a variety of reference systems are in use and with time and experience it is suspected that one of these will be demonstrated to be superior to the others

# CRITERIA FOR NOMENCLATURE OF THE DEFLECTIONS

1 The symbols P $T_P$ QRS T and U are to be used to represent the deflections or groups of deflections encountered in the electrocardiogram Criteria for the use of these symbols apply to all leads unipolar and bipolar normal and abnormal

2 The P wave is normally the gradual initial deflection of any group and may be a summit or a depression The level of reference from which its voltage is measured is the isoelectric level (T P or U P interval) If it displays turning points on either side of its reference level it is described as diphasic If the initial turning point is above this level it is said to be of the plus minus (+—) type and if below it is said to be of the minus plus (—+) type

In esophageal and intracardiac leads the P wave will usually be composed of multiple rapid deflections not unlike the QRS group of leads from the body surface It is recommended that the same symbols and *criteria* of application be used as for the initial ventricular group (see par 5 below) but that the symbol for each atrial deflection be followed by the subscript P e g $Q_P$ $R_P$, $S_P$ $R_P$ $S_P$ $R_P$ $S_P$ Further the level of reference of these deflections as for the P wave itself is to be the isoelectric line

3 The $T_P$ wave may be found in the P R *segment* that part of the trace between the end of the P wave and the beginning of QRS It usually continues through the QRS interval If discernible in body surface leads it is a shallow deflection usually below but sometimes above its level of reference the isoelectric line In esophageal and intracardiac leads it is often of larger amplitude and may be diphasic When there is atrioventricular block an S $T_P$ *segment* preceding and a low summit $U_P$ following the $T_P$ may be identified in such leads

4 In the majority of electrocardiograms the QRS complex is superimposed on the $T_P$ deflection For this reason the level of reference from which the voltage of QRS is measured should be the level at which the first of the QRS components begins The voltage of an upward QRS deflection should be *measured* by estimating the vertical distance between the upper edge of the trace at the beginning of the QRS interval and the upper edge of the trace at the apex of the deflection The voltage of a downward deflection should be determined by

Modified from "The Standardization of Electrocardiographic Nomenclature" by a Committee of the American Heart Association J A M A 121 1347 1943

estimating the vertical distance between the lower edge of the trace at the beginning of the QRS interval and the lower edge of the trace at the bottom of the deflection

5 In order to indicate how the QRS complex should be subdivided for the purpose of assigning symbols to its deflections it should be borne in mind that the first deflection begins at the onset of the QRS interval when the trace first leaves the reference level From this point the trace rises or falls to a turning point where the direction of its motion is reversed It may pass through a second and third turning point or even more causing notches before crossing to the opposite side of the reference level * At this crossing the first deflection ends and the second begins The second deflection necessarily opposite in direction to the first must display one turning point and may display many It does not end until the trace crosses the reference level for the second time There may be a deflection which begins at the second crossing and ends at the S T junction No part of the QRS complex which does not cross the reference level should be considered a separate deflection If the S T junction is displaced in a direction opposite to the turning point of the last deflection of QRS that portion of QRS which lies between this point and the S T junction should be considered as a part of the last deflection

The earliest QRS deflection which lies above the reference level should be labeled R Any downward deflection which precedes R should be labeled Q The first of any downward deflections which may follow R should be labeled S The first of any upward deflections which may follow S should be labeled R′ and the first of any downward deflections which may follow R should be labeled S If it is necessary to label still later deflections of the QRS group the symbols R S and so on should be used in accordance with the same principles When R is absent and the QRS complex consists of a single downward deflection this deflection should be labeled QS In statistical studies QS Q and S deflections should be considered separately For the purpose of roughly indicating the size of the components of the QRS group relative to each other the upper case letters Q R and S may be used for larger deflections and the lower case letters q r and s for smaller deflections

A deflection is notched when it displays more than one turning point on the same side of the reference level A deflection is slurred when it displays a distinct and local thickening on either limb or at its

* When the trace is descending it crosses the reference level at the instant when its lowest margin reaches a position below that which it occupied at the beginning of the QRS interval When the trace is ascending it crosses the reference level at the instant when its upper margin reaches a position above that which it occupied at the beginning of the QRS interval

apex owing to a sudden and pronounced change in the slope of the curve

When the form of the QRS complex varies from moment to moment because of the effect of respiratory movements on the position of the heart or for some similar reason the classification of this complex should be determined by the variety of complex which is most abundant or if no type is numerically predominant by the outline of the complexes which are of intermediate form Small QRS complexes (largest deflection less than 0 5 mv ) which display more than three components or multiple slurring and notching should be classed as small and bizarre or vibratory

6 The term S T *junction* (or J) should be used to *indicate the point* or shoulder which marks the end of the QRS complex the *point where* the steep slopes of the QRS deflections are more or less abruptly replaced by the more gradual slopes which precede or comprise the first limb of the T wave In many electrocardiograms the S T junction is followed by a nearly horizontal or gently sloping segment which lies on above or below the reference level and ends with the onset of a much steeper slope that rises or falls to the apex of T The term S T segment is used for this part of the ventricular complex when it exists even though electrophysiologically it is the earliest part of the T deflection When there is no point between the S T junction and the apex of T at which a sharp change in the slope of the trace occurs this part of the ventricular complex should be called the *first limb of the T wave* When the term S T segment is used *without* reference to some particular electrocardiogram or to some particular class of electrocardiograms it should be understood to refer merely to that part of the ventricular complex which immediately follows the S T junction

The reference level for the measurement of the displacement of the S T *junction* (or J) should be the $T_1$ level The level of reference for the measurement of the S T *segment* the T wave and the U wave should be the isoelectric level when this can be determined otherwise it should be the level of the trace at the beginning of the QRS interval

7 The term diphasic T waves should be applied to those final ventricular deflections which present two distinct turning points one on each side of the level of reference as described for the T waves (see par [illegible] above)

Other terms used in describing the T wave are notched when two apices are noted in the same direction separated by a movement of the curve toward the base line rounded apex when the usual pointed apex is replaced by a curve coved when the ST segment is convex upward but not crossing the line of reference and is followed by an inverted T wave

estimating the vertical distance between the lower edge of the trace at the beginning of the QRS interval and the lower edge of the trace at the bottom of the deflection

5 In order to indicate how the QRS complex should be subdivided for the purpose of assigning symbols to its deflections it should be borne in mind that the first deflection begins at the onset of the QRS interval when the trace first leaves the reference level From this point the trace rises or falls to a turning point where the direction of its motion is reversed It may pass through a second and third turning point or even more causing notches before crossing to the opposite side of the reference level * At this crossing the first deflection ends and the second begins The second deflection necessarily opposite in direction to the first must display one turning point and may display many it does not end until the trace crosses the reference level for the second time There may be a deflection which begins at the second crossing and ends at the S T junction No part of the QRS complex which does not cross the reference level should be considered a separate deflection If the S T junction is displaced in a direction opposite to the turning point of the last deflection of QRS that portion of QRS which lies between this point and the S T junction should be considered as a part of the last deflection

The earliest QRS deflection which lies above the reference level should be labeled R Any downward deflection which precedes R should be labeled Q The first of any downward deflections which may follow R should be labeled S The first of any upward deflections which may follow S should be labeled R′ and the first of any downward deflections which may follow R′ should be labeled S′ If it is necessary to label still later deflections of the QRS group the symbols R″ S″ and so on should be used in accordance with the same principles When R is absent and the QRS complex consists of a single downward deflection this deflection should be labeled QS In statistical studies QS Q and S deflections should be considered separately For the purpose of roughly indicating the size of the components of the QRS group relative to each other the upper case letters Q R and S may be used for larger deflections and the lower case letters q r and s for smaller deflections

A deflection is notched when it displays more than one turning point on the same side of the reference level A deflection is slurred when it displays a distinct and local thickening on either limb or at its

* When the trace is descending it crosses the reference level at the instant when its lowest margin reaches a position below that which it occupied at the beginning of the QRS interval When the trace is ascending it crosses the reference level at the instant when its upper margin reaches a position above that which it occupied at the beginning of the QRS interval

8 INTRAVENTRICULAR CONDUCTION
- 8 0 QRS Interval Normal
- 8 1 Delayed Intrinsicoid (RS) Deflection Left
- 8 2 Delayed Intrinsicoid (RS) Deflection Right
- 8 3 QRS Interval Prolonged
- 8 4 QRS Interval Prolonged Intermittent
- 8 5 Bundle branch Block Left
- 8 6 Bundle branch Block Right
- 8 7 Intraventricular Block Unclassified
- 8 8 Intraventricular Block Intermittent
- 8 9 Anomalous Atrioventricular Excitation

9 ELECTRICAL SYSTOLE
- 9 1 Q T Interval Prolonged

10 ELECTRICAL AXES
- 10 0 No Deviation of the Electrical Axis of QRS
- 10 1 Left Deviation of the Electrical Axis of QRS
- 10 2 Right Deviation of the Electrical Axis of QRS
- 10 3 Left Deviation of the Electrical Axis of T
- 10 4 Right Deviation of the Electrical Axis of T
- 10 5 Deviation of the Electrical Axis of QRST (Ventricular Gradient)
- 10 6 Left Deviation of the Electrical Axis of P
- 10 7 Right Deviation of the Electrical Axis of P

11 DEFLECTIONS
- 11 0 Normal Deflections
- 11 1 High Voltage of P Wave
- 11 2 Low Voltage of P Wave
- 11 3 Broad P Wave
- 11 4 High Voltage of QRS
- 11 5 Low Voltage of QRS
- 11 6 Broad Q Wave
- 11 7 Elevation of S T Junction (J)
- 11 8 Depression of S T Junction (J)
- 11 9 High Voltage of T Wave
- 11 10 Low Voltage of T Wave
- 11 11 Unusual U Wave
- 11 12 Electrical Alternans

12 SPATIAL VECTORCARDIOGRAM
- 12 1 P Form
- 12 2 QRS Form
- 12 3 Junction of QRS Form and T Form
- 12 4 T Form

13 MISCELLANEOUS
- 13 0 Other Conditions Not Listed Above

# CRITERIA FOR INTERPRETATION

## DIAGNOSTIC TITLES

1 Sinus Mechanisms
  - 1 0 Normal Sinus Rhythm
  - 1 1 Sinus Tachycardia
  - 1 2 Sinus Bradycardia
  - 1 3 Sinus Arrhythmia
  - 1 4 Sinus Arrest (Sino atrial Block Sinus Pause)

2 Atrial Mechanisms
  - 2 1 Atrial Premature Systole
  - 2 2 Atrial Tachycardia
  - 2 3 Atrial Flutter
  - 2 4 Atrial Fibrillation
  - 2 5 Wandering Pacemaker

3 Atrioventricular (A V) Nodal Mechanisms
  - 3 1 Atrioventricular (A V) Nodal Premature Systole
  - 3 2 Atrioventricular (A V) Nodal Rhythm
  - 3 3 Atrioventricular (A V) Nodal Tachycardia
  - 3 4 Atrioventricular (A V) Nodal Escape
  - 3 5 Supraventricular Tachycardia

4 Ventricular Mechanisms
  - 4 1 Idioventricular Rhythm
  - 4 2 Ventricular Escape
  - 4 3 Ventricular Premature Systole
  - 4 4 Ventricular Tachycardia
  - 4 5 Ventricular Fibrillation

5 Parasystole
  - 5 1 Atrioventricular (A V) Dissociation with Interference
  - 5 2 Other Parasystolic Rhythms

6 Mechanisms of Undetermined Origin
  - 6 1 Premature Systole of Undetermined Origin
  - 6 2 Tachycardia of Undetermined Origin

7 Atrioventricular (A V) Conduction
  - 7 0 P R Interval Normal
  - 7 1 Incomplete A V Block (Prolonged A V Conduction Time)
  - 7 2 Incomplete A V Block with Dropped Beats
  - 7 3 Complete A V Block

1 1 *Sinus Tachycardia* —This is the same as 1 0 except that the rate exceeds 100 per minute

1 2 *Sinus Bradycardia* —This is the same as 1 0 except that the rate is less than 60 per minute

1 3 *Sinus Arrhythmia* —This is the same as 1 0 except that the P waves followed by the ventricular deflections occur at irregular intervals The arrhythmia is usually phasic and related to respiration (Fig 2) Arbitrarily the duration of cycle lengths must vary by 10 percent or more for the rhythm to be called sinus arrhythmia

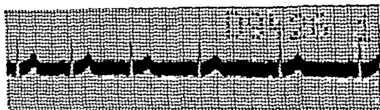


Fig 2

1 4 *Sinus Arrest (Sino-atrial Block Sinus Pause)* —In a sinus rhythm a P wave and its accompanying ventricular deflections fail to appear at the expected time The pause which results is usually slightly shorter than two normal cycles (Fig 3)

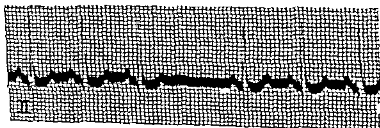



Fig 3

2 Atrial Mechanisms*

2 1 *Atrial Premature Systole* —The premature beat compared with the sinus beats has the following characteristics in at least one lead a P wave different in form a P R interval which is usually as long or longer a ventricular complex which may be the same (Fig 4A) somewhat different (slightly aberrant, Fig 4B) quite different (markedly

*The rubric "... sinus premature systole" has been omitted from this edition because ... pected with a ... al leads now used that the criteria for its diagnosis are ... if e ... met

## 1 Sinus Mechanisms

1.0 *Normal Sinus Rhythm*.—The P wave is upright in Leads I (except in dextrocardia), II, $V_F$, in the anterior and left anterolateral thoracic leads, and in the juxtaventricular and subventricular esophageal leads. *Deviations from this rule may be encountered occasionally* with unusual structure or position of the atria. The P wave occurs regularly at a rate of 60 to 100 per minute (Fig. 1). It is followed by the ventricular deflections QRST except when the ventricular muscle is absolutely refractory or when there is a high grade of atrioventricular block.

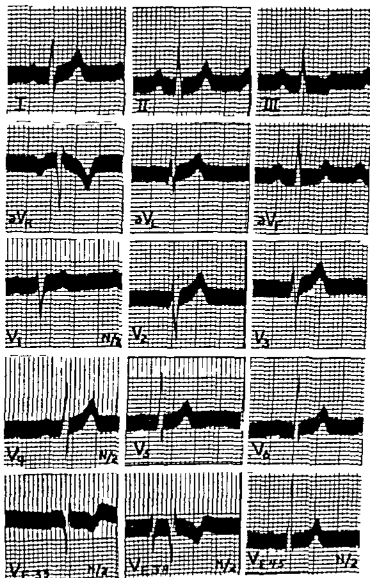



Fig. 1

2 2 *Atrial Tachycardia* —There is a rapid and regular succession of P waves which are different in form from the P waves of the basic sinus rhythm The QRS group may be normal (Fig 6) slightly aberrant or markedly aberrant When it is markedly aberrant differentiation from a ventricular tachycardia is difficult Frequently the P wave coincides with the T wave of the preceding complex thus making differentiation from other supraventricular tachycardias difficult (see 3 5) The atrial deflections in general are seen best in a lead from the right sternal edge (Fig 7) or from the esophagus or from within the atrium Ventricular deflections may be half as frequent as the atrial deflections (1 2 ventricular response Fig 7) In most instances the rate of the atria is between 160 and 280 per minute

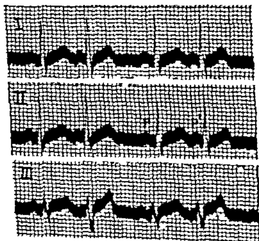



Fig 5

2 3 *Atrial Flutter* —Atrial activity is represented in the electrocardiogram by regular continuous oscillations (P waves) which are uniform except where distorted by ventricular deflections (Fig 8) These oscillations are recorded best in Leads II III and $V_F$ where they are ascribed to an inverted P wave followed by a prominent upright $T_P$ wave They are also recorded well in lead $V_1$ and in juxta atrial leads

aberrant Fig 4C) or absent (blocked atrial premature systole**, Fig 4D) The cycle preceding and the cycle following the premature systole usually have a combined length which is less than two normal cycles When an atrial premature systole regularly follows each normal systole *the electrocardiogram is said to show coupled beats (Fig 5)*

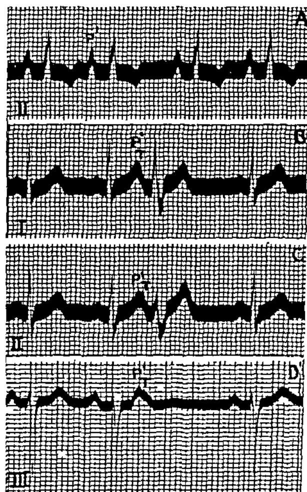



Fig 4

*This term is inaccurate in that the ventricular response is blocked rather than the atrial premature systole Nevertheless it is so generally used that it has been retained

22 *Atrial Tachycardia* —There is a rapid and regular succession of P waves which are different in form from the P waves of the basic sinus rhythm The QRS group may be normal (Fig 6) slightly aberrant or markedly aberrant When it is markedly aberrant differentiation from a ventricular tachycardia is difficult Frequently the P wave coincides with the T wave of the preceding complex thus making differentiation from other supraventricular tachycardias difficult (see 35) The atrial deflections in general are seen best in a lead from the right sternal edge (Fig 7) or from the esophagus or from within the atrium Ventricular deflections may be half as frequent as the atrial deflections (1 2 ventricular response Fig 7) In most instances the rate of the atria is between 160 and 280 per minute

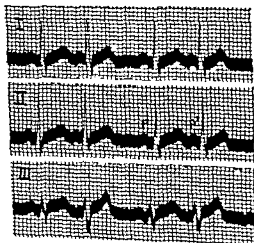



Fig 6

23 *Atrial Flutter* —Atrial activity is represented in the electrocardiogram by regular continuous oscillations (P waves) which are uniform except where distorted by ventricular deflections (*Fig* 8) These oscillations are recorded best in Leads II III and $V_F$ where they are ascribed to an inverted I wave followed by a prominent upright $T_P$ wave They are also recorded well in lead $V_1$ and in juxta atrial leads

aberrant Fig 4C) or absent (blocked atrial premature systole** Fig 4D) The cycle preceding and the cycle following the premature systole usually have a combined length which is less than two normal cycles When an atrial premature systole regularly follows each normal systole the electrocardiogram is said to show coupled beats (Fig 5)

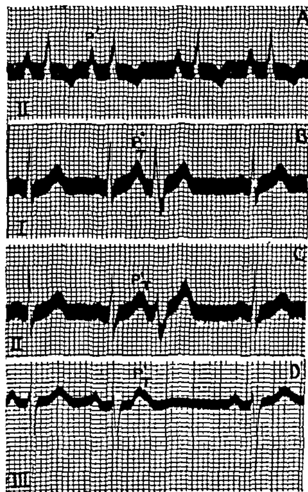



Fig 4

* This term is inaccurate in that the ventricular response is blocked rather than the atrial premature systole Nevertheless it is so generally used that it has been retained

where the P wave may assume a biphasic character They occur at a rate ranging from 200 to 380 per minute The ventricular deflections usually occur regularly at a rate which is slower than and in constant ratio to the atrial rate e g 1 2 1 3 1 4 At times these ratios change in the same tracing The ventricular rhythm is then irregular (Fig 9) Rarely the ratio is 1 1 and such electrocardiograms are usually indistinguishable from other supraventricular tachycardias (see 3 5) *Differentiation* from an atrial tachycardia with a rapid atrial rate and atrioventricular block is often difficult and at times impossible

2 4 *Atrial Fibrillation* —Atrial activity is represented by irregular variable deflections (f waves Fig 10) which have a rate in the neigh

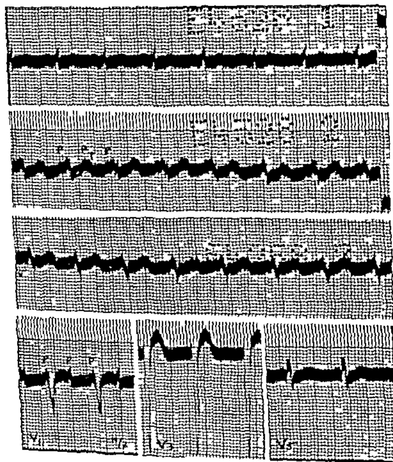
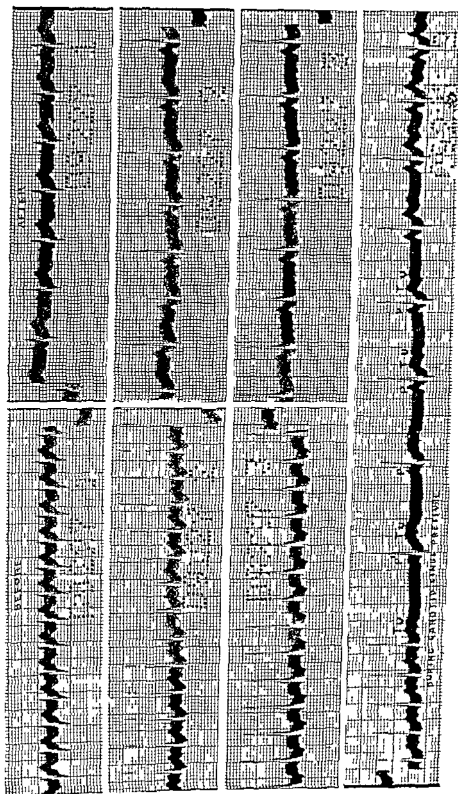



FIG 9


AFTER
BEFORE
DURING CAROTID SINUS PRESSURE

is characteristically inverted in Leads II III and $V_F$ and may occur before during, or after the QRS deflections depending upon whether excitation reaches the atria before during or after it has reached the

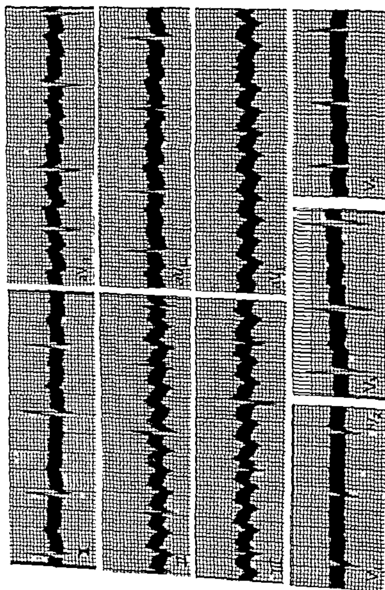


Fig 9

*borhood of 150 per minute* *These are usually recorded best in* esophageal leads in Lead $V_1$ or in intra atrial leads (Fig 11 $V_{R1}$) The ventricular deflections occur at irregular intervals except when there is complete atrioventricular block or when there is a tachycardia arising in a lower center (see 5 2)

2 5 *Wandering Pacemaker* —In a sinus rhythm the pacemaker may change its location within the sino atrial node or from the sino atrial node to the atrioventricular node The former shift is recognized by relatively minor changes in the shape of the P wave the latter by com

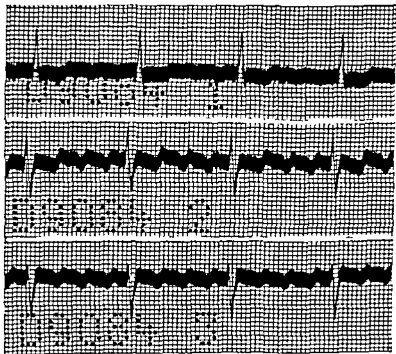


Fig 8

plete inversion of the P wave in Leads II III and $V_F$ and shortening or reversal of the P R interval (Fig 12) When the pacemaker is in the atrioventricular node, the P wave may precede coincide with or follow the QRS complex During the change from a sino atrial to an atrioventricular nodal rhythm there may be several P waves transitional in form (fusion P waves)

3 ATRIOVENTRICULAR (A V) NODAL MECHANISMS

3 1 *Atrioventricular (A V) Nodal Premature Systole* —The P wave

P wave a second QRS may follow the P This phenomenon is called reciprocal rhythm and is ascribed to re entry of the impulse from the A V node to the ventricles

33 *Atrioventricular (A V) Nodal Tachycardia* —Same as 32 except that the rate is over 100 per minute and may be as fast as 270 per minute (Fig 15) In certain records the P waves cannot be recognized and the rhythm cannot be differentiated with certainty from other forms of supraventricular tachycardia In others there is retrograde

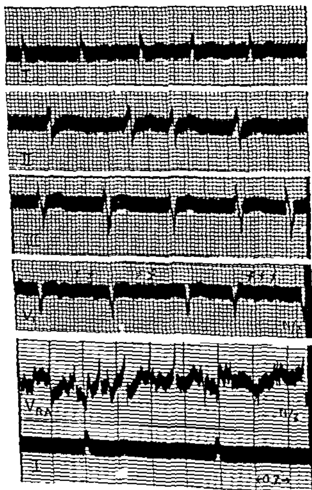



Fig 11

ventricles The ventricular deflections are supraventricular in type but in most instances slightly aberrant *The cycle preceding and the cycle following the premature systole have a total duration usually less than two normal cycles* (Fig 13)

Rarely a premature systole of this type fails to involve the atria because of retrograde block or because the atria are refractory These chambers then continue to respond to a pacemaker at a higher level and the P waves are not altered in shape The combined duration of the cycle preceding and the cycle following *the abnormal beat is equal* to two normal cycles Such beats are indistinguishable from His bundle premature systoles They may be interpolated (see 1 3)

3 2 *Atrioventricular (A V) Nodal Rhythm* —A succession of regular systoles arise from the atrioventricular node at a rate usually between 50 and 80 *per minute* (Fig 14) The electrocardiographic characteristics of each beat may be one of the several described for a premature systole from the A V node (Fig 13) Rarely when QRS precedes the

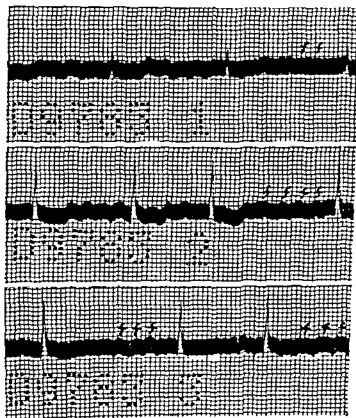


Fig 10

The escaped complex has the features of an A V nodal premature systole except that it is not premature and the ventricular complex usually shows no aberration (Fig 16 A) If the atrial muscle has been

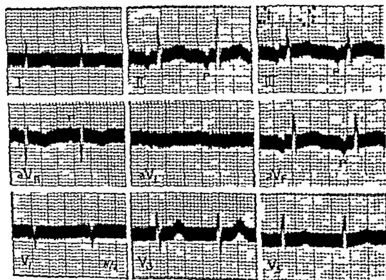



Fig 14

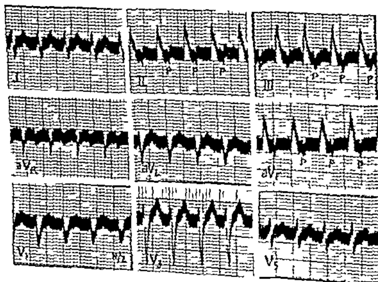



Fig 15

block and the basic atrial rhythm is undisturbed The latter are sometimes called His bundle tachycardias

3 4 *Atrioventricular (A V) Nodal Escape* —A single delayed impulse emanates from the atrioventricular node when the sinoatrial node is depressed or just after a rapid ectopic atrial rhythm comes to an end

Fig 12

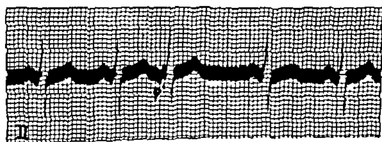


Fig 13

In the latter instance the QRS is markedly aberrant ("ventricular rhythm") but this may occur in the former instance if there is an associated bundle branch block. The rate in either is usually between 30 and 40 beats per minute but may be faster. The atria may show a normal sinus, atrial, atrio ventricular nodal, other rhythm (flutter or fibrillation) or standstill. The atrial deflections bear no fixed relation to the QRS deflections. Idioventricular rhythm is seen most commonly in complete atrioventricular heart block (7 3), less often in certain of the parasystolic rhythms (Fig 17A)




Fig 17

4 2 *Ventricular Escape*—A single impulse emanates from a focus below the atria when there is depression of higher centers. The interval preceding it is longer than the usual cycle lengths of the record. Most frequently this focus is in the common bundle and the electrocardiogram shows a delayed supraventricular QRS with no retrograde P wave because of *retrograde block (see 3 1 and Fig 16B)*. Rarely the focus may be below the *bifurcation and the QRS* is then markedly aberrant (Fig 16C)

4 3 *Ventricular Premature Systole*—The premature beat arises from a center in the ventricles below the bifurcation of the common bundle. The QRS deflections are abnormally wide, notched and slurred and are followed by a T wave which usually is opposite in direction to the main QRS deflections (*markedly aberrant QRS*). Because of retrograd

excited by an impulse from the sino atrial node before the new center escapes or if there is retrograde block the only abnormality will appear as a delayed ventricular complex the normal *P* wave coming before (Fig 16B) with or after the QRS group When this occurs it cannot be said whether the new center is in the A V node or in the common bundle The more inclusive term, ventricular escape is then used to describe it (see 4 2)

3 5 *Supraventricular Tachycardia* —The term supraventricular tachycardia is a collective one which includes all rapid rhythms arising in the sino atrial node atrium A V node or common bundle It is a rhythm with a rate of over 100 per minute and ventricular complexes of supraventricular type but without distinguishable atrial deflections

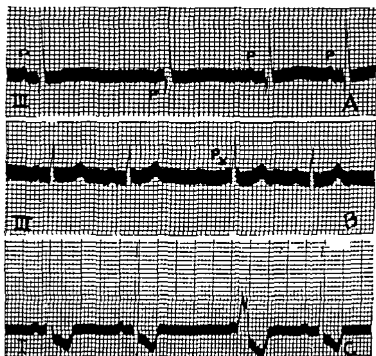



Fig 16

## 4 Ventricular Mechanisms

4 1 *Idioventricular Rhythm* —When the atria are dissociated from the ventricles a pacemaker for the lower chambers may be established in the common bundle* (Fig 17A) or one of its branches (Fig 17B)

*An apparent discrepancy and a real overlap will be noted here between the terms supraventricular (3 5) and idioventricular (4 1) Actually the former term is descriptive of the appearance of QRS only the latter implies a separate pacemaker for the ventricles as compared to the atria This double approach empiric and physiologic is unfortunate but well fixed by usage

from 150 to 250 per minute The rhythm is regular but at times very slight irregularity of the R R intervals may be found The form of the QRS complexes may also vary slightly The atrial rhythm often cannot be recognized (Fig 19) If recognized it may be found to be a normal sinus or other atrial rhythm Sometimes there is retrograde conduction

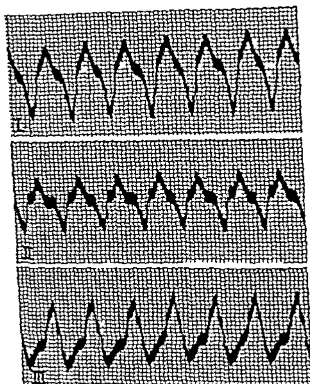



Fig 19

and the atria respond to the ventricles in a 1 1 1 2 or higher ratio In such instances P waves inverted in Leads II III and $V_F$ will be seen at the appropriate intervals In some unusual instances successive ventricular beats are characterized by a principal wave which deflects first in one then in an opposite direction from the baseline Such are said to display bidirectional ventricular tachycardia (Fig 20)

4 5 *Ventricular Fibrillation* — There are large continuous oscillations of the string shadow irregular in form and rate Atrial deflections usually cannot be identified (Fig 21) in leads from the surface of the body

block the rhythm of the atria is not disturbed For this reason the combined length of the cycle preceding and the cycle following the premature systole is equal to two normal cycles and the long pause following this systole is said to be compensatory (Fig 18A) This pause may not be compensatory if retrograde conduction occurs and disturbs the atrial rhythm The pause also may not be compensatory if the atrial

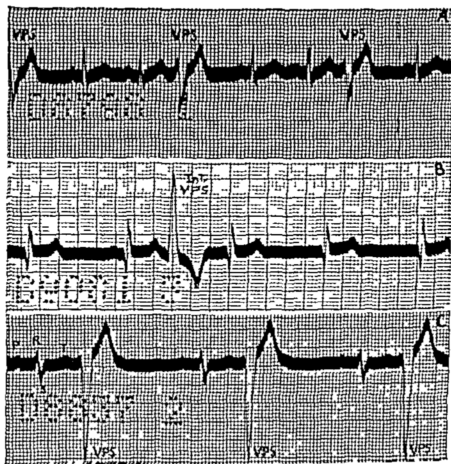



Fig 18

impulse occurring after the premature beat reaches the ventricles when they are no longer refractory from that beat In this event the premature systole is truly an extrasystole or *interpolated premature systole* (Fig 18B) When a ventricular premature systole regularly follows each normal systole the electrocardiogram is said to show coupled beats or coupling (Fig 18C)

1 1 *Ventricular Tachycardia* —A rapid succession of beats each with the characteristics of a ventricular premature systole occurs at rates

from 150 to 250 per minute The rhythm is regular but at times very slight irregularity of the R R intervals may be found The form of the QRS complexes may also vary slightly The atrial rhythm often cannot be recognized (Fig 19) If recognized it may be found to be a normal sinus or other atrial rhythm Sometimes there is retrograde conduction

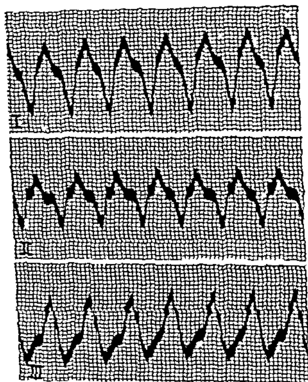



Fig 19

and the atria respond to the ventricles in a 1 1 1 2 or higher ratio In such instances P waves inverted in Leads II III and $V_F$ will be seen at the appropriate intervals In some unusual instances successive ventricular beats are characterized by a principal wave which deflects first in one then in an opposite direction from the baseline Such are said to display bidirectional ventricular tachycardia (Fig 20)

4 5 *Ventricular Fibrillation* —There are large continuous oscillations of the string shadow irregular in form and rate Atrial deflections usually cannot be identified (Fig 21) in leads from the surface of the body

5 PARASYSTOLE

5 1 *Atrioventricular (A V) Dissociation with Interference* —Independent pacemakers control the atria and the ventricles The former show a sinus rhythm the latter an idioventricular (His bundle) rhythm

Fig 20

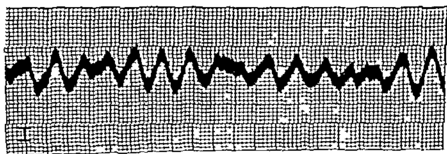


Fig 21

which is faster than the sinus rhythm The P waves bear no relation to the ventricular complexes except in occasional instances In these instances excitation from the atria reaches the ventricles when they are not refractory and after a normal A V conduction time causes a ventricular systole earlier than the one expected from the idioventricular center (Fig 22)

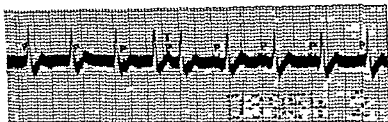


Fig 22

5 2 *Other Parasystolic Rhythms* —All rhythms other than described under 5 1 in which an independent center controls the ventricles either continuously or intermittently in the absence of atrioventricular block

6 Mechanisms of Undetermined Origin

6 1 *Premature Systole of Undetermined Origin* —A premature systole the origin of which cannot be ascertained

6 2 *Tachycardia of Undetermined Origin* —A rhythm with a rate over 100 per minute the origin of which cannot be ascertained

7 Atrioventricular Conduction

7 0 *P R Interval Normal* —The longest* P R interval found in the bipolar or unipolar extremity leads is regarded as indicating the P R interval It is measured from the beginning of the P wave to the beginning of QRS whether this be represented by a Q wave or an R wave With normal heart rates its upper limit in adults is 0 20 sec in adolescents ages 14 to 17 years 0 18 sec and in children under 14 years of age 0 16 sec In precordial leads its maximum duration may be longer

7 1 *Incomplete A V Block (Prolonged A V Conduction Time)* —The P R interval is longer than the maximum defined

* The longest P R interval is not necessarily the correct one but because it does not usually differ from the latter by more than 0.01 sec and because it is easily measured it is preferred The most exact approximation to the true P R interval is obtained by measuring the longest time between the beginning of P and the end of QRS in the [illegible] and [illegible] extremity leads and subtracting from this the longest QRS interval found in these leads

*7 2 Incomplete A V Block with Dropped Beats*—A degree of A V block in which certain P waves of a sinus rhythm are not followed by the ventricular deflections (Fig 23A and Fig 23B) The block may be of various grades depending on the ratio of P waves to the QRS groups Often the P R interval becomes progressively longer in the cycles which precede the dropped ventricular beat (Fig 23A)

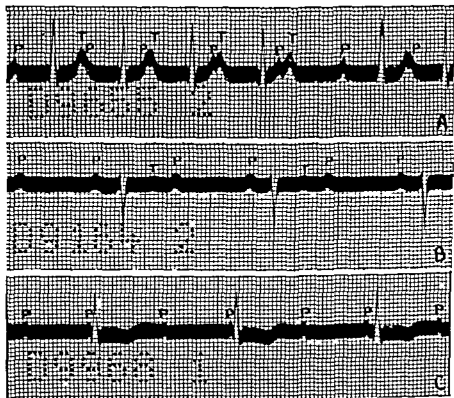



Fig 23

*7 3 Complete A V Block*—A degree of A V block in which no impulses reach the ventricles from the atria The ventricles respond to a pacemaker situated in a center below the block either in the His bundle (Fig 23C) or in the ventricles In the latter event the ventricular complexes are aberrant The ventricles may respond to centers in both locations so that the QRS groups vary in form The rhythm of the ventricles is usually regular and the rate is between 30 and 40 per minute but may be faster Rarely the idioventricular rhythm may be interrupted by a supraventricular impulse when the P wave falls near the end of the ventricular T wave The phenomenon is attributed to a supernormal phase of conduction and may result in occasional retro

grade as well as forward conduction through the junctional tissues in the presence of A-V block.

8. INTRAVENTRICULAR CONDUCTION

8.0. *QRS Interval Normal.*—The longest QRS interval found in the bipolar or unipolar extremity leads is regarded as most nearly correct. It is measured from the beginning (Q or R) to the end of the QRS group. Its upper limit is 0.10 sec. in adults, 0.09 sec. in children from 5 to 14 years of age, 0.08 sec. in children under 5 years. In precordial leads its maximum duration is slightly longer.

8.1. Delayed Intrinsicoid (RS) *Deflection Left.*—In leads from the left side of the precordium ($V_5$ and $V_6$) the time measured from the beginning of QRS to the peak of the R wave* exceeds 0.05 sec.

8.2. *Delayed Intrinsicoid (RS or RS′) Deflection Right.*—In leads from the right side of the precordium ($V_1$ and $V_2$) the time measured from the beginning of QRS to the peak of the R wave exceeds 0.03 sec. When there is a second R wave (R′) in these leads, the peak of this wave is used as the point of measurement if RS′ exceeds RS in size.

8.3. *QRS Interval Prolonged.*—The QRS interval is between 0.10 and 0.12 sec. Such prolongation may be caused by hypertrophy, by incomplete block of one or the other bundle branches, by both, or by unknown factors.

Many records will not display features of QRS sufficiently characteristic to indicate which of these reasons underlies the prolonged QRS interval. It is suggested that all these curves be classified simply as QRS interval prolonged. Further, it is likely that incomplete block of the right bundle branch and probably of the left bundle branch can exist with a QRS interval of 0.1 sec. or less. When either is suspected, it is suggested that the curve be classified for the present under category 13.0 (Miscellaneous).

8.4. *QRS Interval Prolonged Intermittent.*—The term is applied to those electrocardiograms displaying a normal sinus or other regular supraventricular rhythm, a constant P-R interval, and QRS groups which are normal and prolonged, as a rule alternately, occasionally irregularly or progressively. When the basic rhythm is atrial fibrillation the QRS interval may be normal in length only after a long diastolic pause.

8.5. *Bundle-branch Block, Left.*—The QRS interval is 0.12 sec. or

* The peak of the R wave represents the beginning of the intrinsicoid deflection and is measured because this is clinically expedient. It is clear that it is produced by the beginning of depolarization of underlying epicardial muscle rather than by its completion. Ordinarily the intrinsicoid deflection is the steepest and largest downstroke in the QRS group, but in some records two deflections of similar character may be found.

more and the components of QRS are notched and slurred In Leads I, $V_L$ sometimes in $V_F$ and in leads from the extreme left side of the precordium and left side of the thorax the initial deflection is usually an R wave which is notched or slurred In these same leads the peak of the R wave or one of its prominent notches occurs relatively late in the QRS interval The S T segment is most often displaced in a direction opposite to the principal QRS deflection and the T wave usually also points in this direction The appearance of QRS in other leads depends principally on the average direction of the electrical axis of QRS (Figs 21 25)

It is fairly certain that hypertrophy of the left ventricle of itself occasionally can cause a QRS interval of 0 12 sec or more These records will differ from the usual ones due to left bundle branch block in that Q waves will be present in Lead I and in the leads from the left arm and from the left side of the precordium and the QRS deflections of the bipolar extremity leads usually will be quite high and relatively free from notching and slurring (see 8 7)

8 6 *Bundle branch Block Right* — The QRS interval is 0 12 sec or

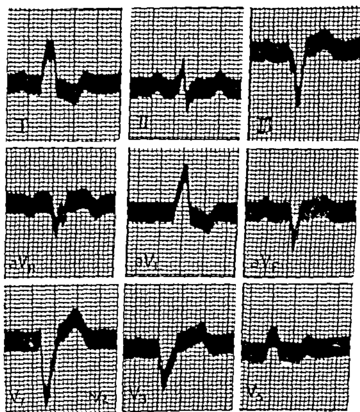
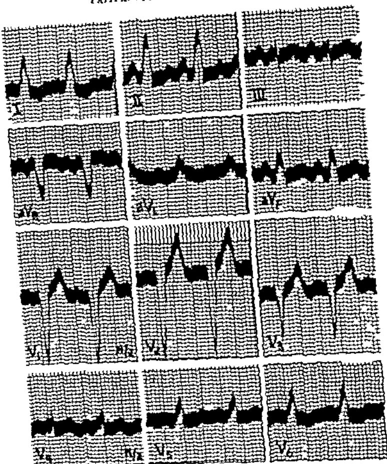



FIG 21




Fi 9

over. The characteristic finding is the occurrence of a late, usually large R or R' in Lead $V_1$, sometimes in Leads $V_2$ and $V_3$, occasionally in leads farther to the *right* ($V_{3R}$, $V_{4R}$). Preceding this there is usually a small r. A record consisting of a deep Q and a broad, slurred R wave is often obtained in *Lead* $V_6$. *Usually* in Lead I and in leads from the left side of the precordium *there is* a narrow R wave of variable size and a broad, slurred S wave.

The electrical axis of QRS, *chamber* hypertrophy, and other factors determine a variety of types which are recognized by the characteristics of the ventricular *complex in the bipolar* and unipolar extremity leads (Figs. 26 and 27). An infrequent *form* is one which resembles left bundle branch block in these leads except for the presence of a Q wave or S wave of variable dimens*ions in* Leads I and $V_L$ (Fig. 28).

more and the components of QRS are notched and slurred In Leads I $V_L$, sometimes in $V_F$ and in leads from the extreme left side of the precordium and left side of the thorax the initial deflection is usually an R wave which is notched or slurred In these same leads the peak of the R wave or one of its prominent notches occurs relatively late in the QRS interval The S T segment is most often displaced in a direction opposite to the principal QRS deflection and the T wave usually also points in this direction The appearance of QRS in other leads depends principally on the average direction of the electrical axis of QRS (Figs 21 25)

It is fairly certain that hypertrophy of the left ventricle of itself occasionally can cause a QRS interval of 0 12 sec or more These records will differ from the usual ones due to left bundle branch block in that Q waves will be present in Lead I and in the leads from the left arm and from the left side of the precordium and the QRS deflections of the bipolar extremity leads usually will be quite high and relatively free from notching and slurring (see 8 7)

8 6 *Bundle branch Block Right* —The QRS interval is 0 12 sec or

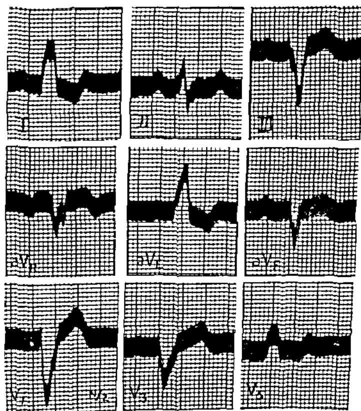



Fig 21

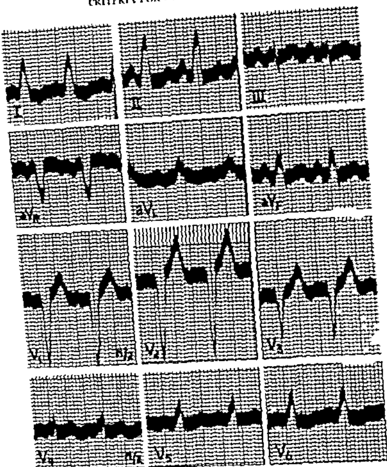



Fig [illegible]

over The characteristic finding is the occurrence of a late usually large R or R' in Lead $V_1$ sometimes in Leads $V_2$ and $V_3$ occasionally in leads farther to the right ($V_{3R}$ $V_{4R}$) Preceding this there is usually a small rS A record consisting of a deep Q and a broad slurred R wave is often obtained in Lead $V_R$ *Usually in Lead I and in leads from the left side of the precordium there is a narrow R wave of variable size and a broad slurred S wave*

The electrical axis of QRS chamber hypertrophy and other factors determine a variety of types which are recognized by the characteristics of the ventricular complex in the bipolar and unipolar extremity leads (Figs 26 and 27) An infrequent form is one which resembles left bundle branch block in these leads except for the presence of a Q wave or S wave of variable dimensions in Leads I and $V_1$ (Fig 28)

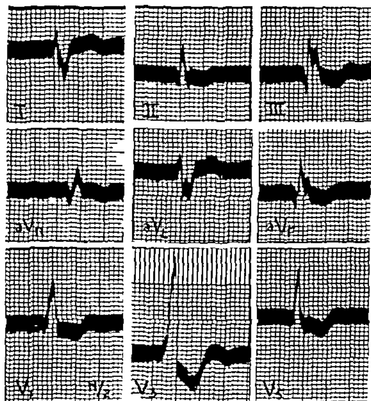



Fig 26

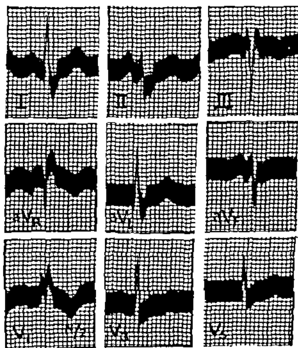



Fig 27

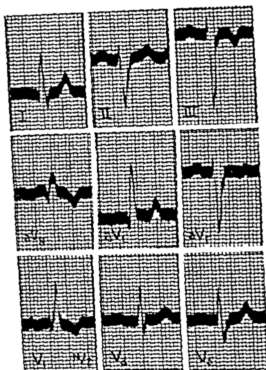



FIG 28

8 *Intraventricular Block Unclassified*—This group includes all electrocardiograms with a QRS interval of 0 12 sec or more not fitting into any of the above classes It is suggested that records with a wide QRS interval ascribed to hypertrophy alone and records variously described as showing arborization block focal block and other types of block be placed in this category until the mechanisms involved in each are clarified

88 *Intraventricular Block Intermittent*—The term is applied to those electrocardiograms displaying a normal sinus or other regular supraventricular rhythm a constant P R interval and QRS groups which are variably normal and abnormal (0 12 sec or more) in width This usually occurs alternately (Fig 29A) but occasionally irregularly (Fig 29b) or progressively When the basic rhythm is atrial fibrillation the QRS interval may be normal in length only after a long diastolic pause This term is intended to include intermittent bundle branch block

8 9 *Anomalous Atrioventricular Excitation** —The record is characterized in one or several of the bipolar and unipolar extremity leads by a slurring or notching of the earliest QRS deflection This additional

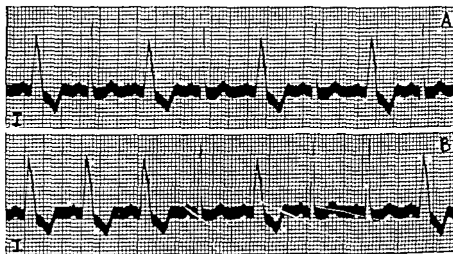



Fig 29

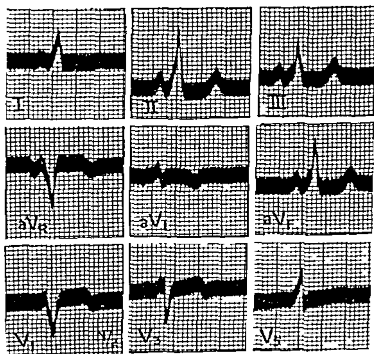



Fig 30

*Although this type of electrocardiogram is not believed to be the result of defective intraventricular conduction it is placed under this category because of the considerable broadening of the *QRS interval* which is present

component of QRS (anomalous component delta wave) is caused by a premature excitation of some part of the ventricular muscle in an anomalous fashion By virtue of this component's prematurity the P R segment appears to be shortened or absent the P R interval abbreviated and the QRS interval prolonged (Fig 30)

The direction of excitation and the location of the anomalously excited portion of the ventricle determine whether the principal QRS deflection particularly in leads from the right side of the precordium will be positive or negative

Records are often seen in which both anomalous and normal QRS complexes occur alternately or intermittently (Fig 31)

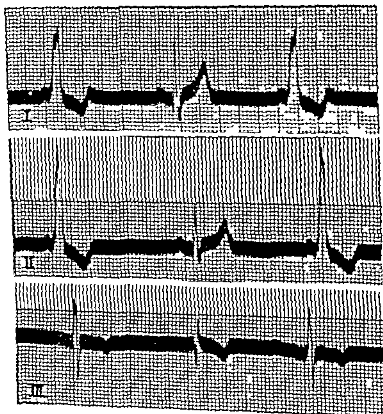



Fig 31

9 Electrical Systole

9 1 *Q T Interval Prolonged* — The Q T interval is measured from the beginning of QRS to the end of the T wave The largest interval

8 9 *Anomalous Atrioventricular Excitation** —The record is characterized in one or several of the bipolar and unipolar extremity leads by a slurring or notching of the earliest QRS deflection This additional

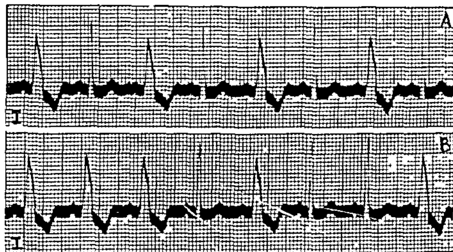



Fig 29

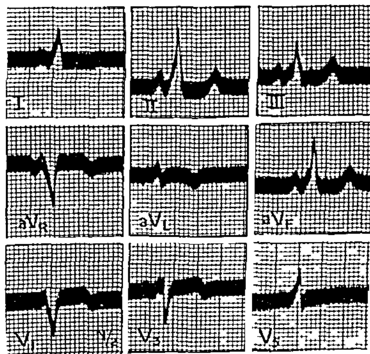



Fig 30

*Although this type of electrocardiogram is not believed to be the result of defective intraventricular conduction it is placed under this category because of the considerable broadening of the QRS interval which is present

positive in Lead $V_R$ and zero in Lead I The definition delimits an angle alpha of +29 to −90 (Figs 32B and C) Usually with this direction in the frontal plane the axis of QRS is deviated well backward as manifested by a shift of the transitional zone to the left

102 *Right Deviation of the Electrical Axis of QRS* —The algebraic sum of the areas of Q R and S in Leads I $V_R$ and $V_L$ is negative

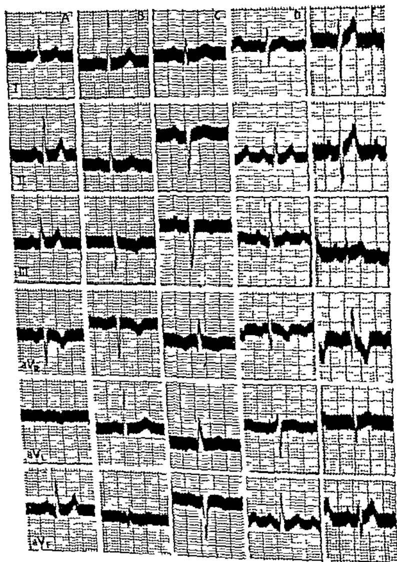



Fig 32

found in any lead from the body surface is regarded as most nearly correct It varies with the rate of the heart beat *A correction for rate* can be made by the easily used formula $Q\text{-}T = K \sqrt{\text{cycle length}}$ The Q-T interval may be regarded as prolonged if K exceeds 0 425

10 Electrical Axis*

10 0 *No Deviation of the Electrical Axis of QRS* —By regarding summits as positive and depressions as negative the algebraic sum of the areas of the QRS deflections of the three bipolar leads and of Lead $V_F$ is positive of Lead $V_R$ negative and of Lead $V_L$ negative zero or positive If this value is zero in Lead I or zero in Lead III the record is also regarded as showing no deviation of the electrical axis provided that the value is positive in the remaining two bipolar extremity leads This is equivalent to an angle alpha* of $+30°$ to $+90°$ (Fig 32A)

Usually the electrical axis of QRS is directed not only downward and to the left but backward The backward direction manifests itself by a transitional QRS ** in a precordial lead to the left of the midline anteriorly The area of QRS deflections in anterior precordial leads to the right of the transitional QRS is negative and to its left positive

In the frontal plane the average size of the electrical axis of QRS is 22 microvolt seconds ($\mu$vs) in adults 17 $\mu$vs in children under 15 years The area of the deflection may be estimated by inspection but can be measured more precisely if desired

10 1 *Left Deviation of the Electrical Axis of QRS* —The algebraic sum of the areas of Q, R and S is positive in Leads I and $V_L$, and negative in Lead III It is negative in Leads $V_F$ and II as well if the degree of deviation is marked*** With extreme deviation this value is

* The term electrical axis refers to the mean manifest electrical potential in space responsible for a designated deflection or deflections Since potential has both size and direction it may be treated as a vectorial quantity Because it occurs in space it has frontal sagittal and transverse components Its length is measured usually in microvolt seconds ($\mu$vs) its direction in terms of an angle it makes with one of the orthogonal coordinates Most of the criteria are concerned with its frontal projection and the angle this makes with the horizontal (angle alpha) available data on its sagittal and transverse components are inadequate for quantitative criteria at this time In the definitions given in rubrics 10 0 10 1 and 10.2 the smaller polar angle between the limits of the angle alpha given is the one that applies

** A transitional QRS in the precordial leads is defined as one having combined characteristics of leads recorded on either side of it The algebraic sum of the areas of its deflections is close to zero

*** The Committee is aware that the quadrants of the rectilinear coordinates in the frontal plane are mislabeled in sign contrasted to mathematical custom It is aware too that deviation to the left is maximal when the angle alpha is 0 However the electrocardiographic custom of regarding records with an angle alpha between 0 and −90 (mathematical quadrant I) as displaying moderate to marked left deviation of the axis is so universal and ingrained that a change at this time does not seem desirable

TABLE 1.—NORMAL CHILDREN NEWBORN TO 1 YEAR OLD SUPINE. SIZE OF THE ELECTROCARDIOGRAPHIC DEFLECTIONS IN THE BIPOLAR EXTREMITY, AUGMENTED UNIPOLAR EXTREMITY AND PRECORDIAL LEADS IS GIVEN IN TENTHS OF A MILLIVOLT

| Lead | P | | | | Q | | | | R | | | | S | | | | RS | | | | ST | | | | T | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | N C | M | M | M | C | M | M | M | N Cw | M | M | M | N Cw | M | M | Mw | N Cases | Mi | M | M | C | M | M | M | No. | M | M | M |
| I | 158 | −1.0 | 2.5 | 0.94 | 164 | 0 | 3.0 | 1.15 | 165 | 0 | 17.0 | 5.58 | 165 | 0 | 15.0 | 4.34 | — | — | — | — | 195 | −0.1 | 0.1 | — | 145 | −2.0 | 6.0 | 3 |
| II | 159 | 0 | 2.5 | 1.63 | 165 | 0 | 5.5 | 1.95 | 165 | 1.0 | 25.0 | 10.04 | 165 | 0 | 8.5 | 2.99 | — | — | — | — | 197 | −0.1 | 0 | — | 145 | 0 | 7.0 | 2.84 |
| III | 158 | −1.0 | 2.0 | 0.80 | 165 | 0 | 9.0 | 3.96 | 164 | 1.0 | 24.0 | 8.91 | 161 | 0 | 6.5 | 2.18 | — | — | — | — | 197 | −0 | 0.2 | — | 145 | −5.0 | 5.0 | 0.75 |
| aVR | 159 | −2.5 | 1.5 | 1.09 | 165 | 0 | 15.0 | 5.92 | 165 | 0 | 9.0 | 2.32 | 165 | 0 | 18.0 | 7.45 | — | — | — | — | 197 | −0.1 | 0.1 | — | 145 | −3.0 | 2.0 | −2.39 |
| aVL | 158 | −1.5 | 1.5 | 0 | 164 | 0 | 5.5 | 1.34 | 165 | 0 | 10.0 | 5.35 | 165 | 0 | 16.0 | 5.[illegible] | — | — | — | — | 197 | −0.1 | 0.1 | — | 145 | −1.5 | 3.5 | 0.97 |
| aVF | 158 | −1.0 | 3.0 | 1.17 | 165 | 0 | 6.0 | 2.36 | 165 | 1.5 | 21.5 | 8.15 | 165 | 0 | 7.5 | 2.29 | — | — | — | — | 197 | −0.05 | 0.2 | — | 145 | −2.0 | 5.0 | 1.68 |
| $V_1$ | 155 | −1.0 | 2.5 | 0.69 | 186 | 0 | 0 | 0 | 137 | 3.0 | 99.0 | 15.61 | 1 3 | 0 | 8.0 | 8.57 | 142 | 3.0 | 50.0 | 22.75 | 161 | −0.2 | 0.1 | — | 1 6 | −8.0 | 6.0 | −.85 |
| $V_2$ | 159 | 0 | 2.5 | 1.13 | 186 | 0 | 0 | 0 | 137 | 3.0 | 43.0 | 19.98 | 121 | 1.0 | 42.0 | 18.35 | 16 | 5.0 | 66.0 | 40.35 | 159 | −0.2 | 0.2 | — | 1?6 | −9.0 | 9.0 | −.84 |
| $V_3$ | 115 | 0 | 3.0 | 1.43 | 117 | 0 | 3.0 | 1.09 | 114 | 3.5 | 40.0 | 0.49 | 112 | 1.0 | 39.0 | 17.3 | 116 | (18.0) | 75.0 | 38.71 | 116 | −0.2 | 0.25 | — | 116 | −7.0 | 6.0 | −1.57 |
| $V_4$ | 142 | 0 | 2.0 | 1.05 | 161 | 0 | 3.0 | 1.52 | 159 | 5.0 | 37.0 | 21.21 | 125 | 0 | 42.0 | 1.88 | 146 | 4.0 | 63.0 | 31.75 | 157 | −0.1 | 0.2 | — | 123 | −7.0 | 6.5 | .46 |
| $V_5$ | 122 | 0 | 0 | 1.14 | 1?2 | 0 | 5.5 | 2.09 | 118 | 5.0 | 51.0 | 14.49 | 119 | 0 | 30.0 | 7.00 | 1 1 | 5.0 | 48.0 | 21.89 | 121 | — | 0.2 | — | 1?2 | −4.0 | 7.0 | 2.98 |
| $V_6$ | 141 | 0 | 2.0 | 0.84 | 142 | 0 | 5.0 | 1.64 | 119 | 0 | 21.0 | 4.00 | 139 | 0 | 30.0 | 2.7 | 161 | 2.0 | 34.0 | 12.17 | 161 | — | 0.1 | — | 1 5 | −3.0 | 6.0 | 2.27 |

B ed on rmal se es re d ed by Ziegl r
6 m th –1 year ge gro p ly

With a greater degree of deviation to the right this value may also be negative in Leads II III and $V_F$ and positive in Lead $V_R$ The definition delimits an angle alpha of +91° to −91° (Figs 32D and E) In the sagittal plane the axis may be directed backward or forward the distinction being made on the appearance of the thoracic leads

10 3 *Left Deviation of the Electrical Axis of T* —The angle alpha of the electrical axis of T is between +29° and −90° The angle is determined from the area and direction of the T wave in various leads as described for QRS in 10 1

10 4 *Right Deviation of the Electrical Axis of T* —The angle alpha of the electrical axis of T is between +91° and −91° The angle is determined from the area and direction of the T wave in various leads as described for QRS in 10 2

10 5 *Deviation of the Electrical Axis of QRST (Ventricular Gradient)* —The electrical axis of QRST is treated as a vector sum of the electrical axes of QRS and T Its mean length in the frontal plane in terms of the area of the ventricular deflections is approximately 16 microvolt seconds (µvs) Its direction in the frontal plane is determined in the normal subject by the direction of the electrical axis of QRS from which it usually does not differ by more than 30 in either direction When the difference between the two is greater than 30 deviation of the ventricular gradient is said to exist

10 6 *Left Deviation of the Electrical Axis of P* —The angle alpha of the electrical axis of P is between +29 and −90 The angle is determined from the area and the direction of the P wave in various leads as described for QRS in 10 1

10 7 *Right Deviation of the Electrical Axis of P* —The angle alpha of the electrical axis of P is between +91 and −91 The angle is determined from the area and direction of the P wave in various leads as described for QRS in 10 2

11 DEFLECTIONS*

11 0 *Normal Deflections* —The range of normal values of atrial and ventricular deflections as *determined in normal subjects in the age* groups newborn to 1 year 1 year to 10 years 10 years to 20 years and 20 years and over are shown in Tables I II III and IV

11 1 *High Voltage of P Wave* —The voltage of the P wave measured in one direction from the isoelectric level exceeds 0 3 mv in any of the

*For practical reasons the criteria for some of these definitions are somewhat arbitrary Therefore the definitions may include values which are at variance with the range shown in the tables of normal values

TABLE III.—NORMAL ADOLESCENTS 10 TO 20 YEARS OLD. SUMMARY OF THE ELECTROCARDIOGRAPHIC DEFLECTIONS IN THE BIPOLAR EXTREMITY, AUGMENTED UNIPOLAR EXTREMITY AND PRECORDIAL LEADS IS GIVEN IN TENTHS OF A MILLIVOLT

| Lead | P | | | | Q | | | R | | | S | | | T | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | N C | M | M | M | M | M | M | M | M | M | M | M | M | M | M | M |
| I | 1 1 | 0.2 | 15 | 0 71 | 0 | 25 | 0 30 | 13 | 150 | 5.[illegible] | 0 | 68 | 1 10 | 02 | 50 | 2 32 |
| II | 191 | 0 | 21 | 1 01 | 0 | 28 | 0 37 | 23 | 200 | 911 | 0 | 63 | 1 9 | 0.2 | 65 | 2 89 |
| III | 1 1 | –1 0 | 1 8 | 0 13 | 0 | 4 6 | 0.50 | 07 | 15 8 | 6 03 | 0 | 90 | 1 18 | –1 9 | 3 9 | 0 65 |
| aV | –14 | –1 75 | –0.2 | –1 03 | 0 | 13 7 | 2 69 | 0 | 8 0 | 1 39 | 0 | 17 0 | 1 91 | –5.2 | –0 1 | –2.36 |
| aV$_L$ | 211 | –1 0 | 1 1 | 0.29 | 0 | 1.2 | 0 31 | 0 | 10 1 | 2.21 | 0 | 11.2 | 2 51 | –2.5 | 5 6 | 0 87 |
| aV$_F$ | 211 | –0 8 | 2.2 | 0 1 | 0 | 3 8 | 0 12 | 1 0 | 21 0 | 8.2 | 0 | 1 9 | 0 99 | –0.5 | 5 4 | 1 76 |
| V$_1$ | 158 | –0.2 | 2.2 | 0 17 | 0 | 1.5 | 0 01 | 0 1 | 16 7 | 5.29 | 0 | 26 6 | 11 99 | –3 5 | 7.5 | 0 22 |
| V$_2$ | 159 | –0.2 | 2 0 | 0 8 | 0 | 0 | 0 | 0.5 | 25.5 | 9 16 | 2 6 | 15 5 | 15 81 | –3 8 | 11 1 | 2 19 |
| V$_3$ | 158 | 0 | 1 8 | 0 13 | 0 | 0 7 | 0 01 | 1 6 | 20 0 | 10 1 | 0 9 | 31 1 | 12 6 | –3 7 | 13.5 | 3 39 |
| V$_4$ | 159 | 0 | 1.5 | 0 6 | 0 | 1 1 | 0 21 | 3 1 | 31 0 | 15 09 | 0 7 | 22 2 | 8.27 | –2 8 | 12 6 | 1 13 |
| V$_5$ | 159 | 0 | 1.2 | 0 55 | 0 | 3 1 | 0 19 | 1.2 | 29 0 | 15 6 | 0 | 1 1 | 3 03 | 0 1 | 10.5 | 3 71 |
| V$_6$ | 158 | 0 | 1 0 | 0 0 | 0 | 4 2 | 0.66 | 3 5 | 2 0 | 11 11 | 0 | 11 3 | 1 71 | 0 | 7 8 | 3 01 |
| V$_7$ | 51 | 0.2 | 0 8 | 0 1 | 0 | 2.5 | 0 9 | 5 0 | 10 0 | 15 0 | 0 | 5 4 | 0 3 | 0 7 | 5 0 | 2 1 |
| V$_{3}$ | 21 | — | 1 9 | 0 7 | 0 | 0 | 0 | 1 5 | 8 0 | 3 8 | 0 | 10 0 | 7 6 | –3 6 | 1 8 | –1 1 |
| V$_4$ | 21 | — | 1 0 | 0 1 | 0 | 1.5 | — | 0 7 | 5 | 2.3 | 1 0 | 9 5 | 5 1 | –2 1 | 1 1 | –1 1 |

This table is based on normal series studied by Sapiero, Limon and Limon, Switzer and Besoain, Kneele de Melo, San Jose and Katz as per

TABLE 11 —NORMAL CHILDREN 1 TO 10 YEARS OLD SUPINE SIZE OF THE ELECTROCARDIOGRAPHIC DEFLECTIONS IN THE BIPOLAR EXTREMITY AUGMENTED UNIPOLAR EXTREMITY AND PRECORDIAL LEADS IS GIVEN IN TENTHS OF A MILLIVOLT

| Lead | P | | | | Q | | | | R | | | | S | | | | RS | | | | S T | | | | T | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | No. Cases | M | M | M | N Cases | M | M | M | N Cases | M | M | M | N Cases | M | M | M | N Cases | M | M | M | N Cases | M | M | M | No. Cases | M | M | M |
| I | 5 | 0.5 | 2.5 | 1.01 | ?7 | 0 | 2.5 | 1.11 | 2?3 | 1.5 | 17.0 | 6.75 | 2?5 | 0 | 8.0 | 88 | — | — | — | — | 297 | −1.0 | 1.0 | — | 223 | 1.0 | 6.5 | 2.53 |
| II | 21 | 0 | 3.0 | 1.63 | 2?8 | 0 | 5.4 | 1.5 | 2 3 | ?0 | ?8.0 | 12.21 | ??3 | 0 | 6.5 | ?38 | — | — | — | — | 297 | −1.0 | 2.0 | — | ?23 | 0 | 7.0 | 3.25 |
| III | ?21 | −1.5 | ?0 | 0.62 | 7 | 0 | 8.0 | ?50 | 23 | 1.0 | 3.0 | 7.99 | ??3 | 0 | 9.0 | 1 0 | — | — | — | — | ?98 | −1.0 | 1.0 | — | ?23 | −3.0 | 4.0 | 0.52 |
| aV | ?93 | −2.0 | 0 | −1.56 | 317 | 0 | 11.0 | 5.70 | 312 | 0 | 6.5 | 1.60 | 31? | 0 | 19.5 | 7 7 | — | — | — | — | ??7 | −1.0 | 1.0 | — | 292 | −6.0 | −0.2 | −2.79 |
| aV | 93 | −1.0 | 2.0 | 0.3 | 317 | 0 | 4.0 | 0.81 | 312 | 0 | 11.8 | 3.15 | 31 | 0 | 11.0 | 3 2 | — | — | — | — | ??7 | −1.0 | 1.0 | — | ?92 | −4.0 | 4.0 | 1.10 |
| aV | ?93 | −0.6 | 2.0 | 1.03 | 317 | 0 | 5.0 | 1.33 | 312 | 0.5 | 21.0 | 9.30 | 31 | 0 | 14.0 | 1.61 | — | — | — | — | ??7 | −1.0 | 1.0 | — | 92 | −1.0 | 6.0 | 1.84 |
| $V_1$ | 198 | −1.0 | 2.5 | 0 2 | 03 | 0 | 0 | 0 | 198 | 0.4 | ?0.0 | 7.15 | 197 | 0 | 36.? | 11.0? | 103 | 6.5 | 46.5 | 8.8 | 131 | −1.0 | 2.0 | — | 197 | −6.0 | 3.0 | −2.86 |
| $V_2$ | 01 | −0.6 | 2.0 | 0.9 | 01 | 0 | 0 | 0 | 19 | ?0 | 8.0 | 1 90 | 196 | 3.0 | 41.0 | 18.31 | 101 | 9.0 | 61.0 | 37.45 | 133 | −1.0 | 2.5 | — | 197 | −8.5 | 6.0 | −1.91 |
| $V_3$ | 18? | 0.1 | 2.0 | 0.80 | 18 | 0 | 1.0 | 0.44 | 181 | 4.0 | 4 0 | 14.00 | 180 | 0 | 36.0 | 13.66 | 87 | 14.0 | 60.0 | 3 17 | 118 | 0 | ?5 | — | 191 | −13.0 | 12.5 | 1.30 |
| $V_4$ | ?00 | 0 | 1.5 | 0.73 | 03 | 0 | 6.0 | 1.?1 | 199 | 4.0 | 50.0 | ?0.16 | 199 | 0 | ?6.0 | 7.53 | 102 | 13.0 | 61.0 | 3 91 | 133 | 0 | ?0 | — | 199 | −4.0 | 14.5 | 4.54 |
| $V_5$ | 198 | 0 | 1.5 | 0.? | 190 | 0 | 8.0 | 1.53 | 186 | 6.0 | 40.0 | 18.97 | 196 | 0 | 15.0 | 3.6 | 88 | 15.0 | 47.0 | 23.89 | 83 | −1.0 | 1.2 | — | 186 | 0 | 12.0 | 4.51 |
| $V_6$ | 197 | 0 | 1.5 | 0.61 | ?01 | 0 | 5.0 | 1.47 | 196 | 5.0 | 2 ?0 | 11.94 | 19 | 0 | 13.0 | 1.?3 | 10? | 5.0 | 26.0 | 14.74 | 93 | −1.0 | 1.0 | — | 196 | 0 | 8.5 | 3.41 |
| $V_7$ | 8 | 0 | 1.0 | 0.50 | ?8 | 0 | 3.0 | 0 8 | ?8 | 5.? | 20.0 | 11.13 | 8 | 0 | 5.5 | 1.?9 | — | — | — | — | 28 | 0 | 0.8 | — | 28 | 0.7 | 6.5 | 3.50 |
| $V_3$ | 8 | −0.8 | 1.5 | 0.57 | 8 | 0 | 0 | 0.09 | 8 | 1.3 | 9.9 | 4.18 | 28 | 0 | 19.5 | 5.51 | — | — | — | — | ?8 | 0 | 0.6 | — | 8 | −4.8 | 0.9 | −1.77 |
| $V_4$ | 8 | −0.4 | 1.5 | 0.44 | 8 | 0 | 2.0 | 0.09 | 8 | 1.0 | 4.8 | 2.80 | 28 | 0 | 10.5 | 8.67 | — | — | — | — | ?4 | 0 | 0.4 | — | 8 | −4.9 | −1.0 | −1.59 |

This table is based on normal series studied by Z egl Sw t l Beso K eese l M lo d Yu Joos nd K mpes

bipolar extremity leads (I II III) or 0 25 mv in any of the precordial leads ($V_1$ to $V_6$) or augmented unipolar extremity leads ($aV_R$ $aV_L$ $aV_F$) or 0 2 mv in any of the unipolar extremity leads ($V_R$ $V_L$ $V_F$)

11 2 *Low Voltage of P Wave* —The voltage of the P wave measured in one direction from the isoelectric level is less than 0 05 mv in all of the commonly recorded surface leads or in all those available

11 3 *Broad P Wave* —The width of the P wave measured at its base exceeds 0 10 sec in the bipolar or unipolar extremity leads A P wave with a duration of 0 12 sec or more in these leads is regarded by some as evidence of intra auricular block

11 4 *High Voltage of QRS* —The voltage is regarded as high when the largest QRS deflection on one side of the reference level (end of the P R segment) exceeds 2 5 mv in any of the bipolar extremity leads or 1 5 mv in any of the unipolar extremity leads or 2 0 mv in any of the augmented unipolar extremity leads or 5 0 mv in any of the usually recorded six precordial leads

11 5 *Low Voltage of QRS* —The voltage is regarded as low when the largest QRS deflection on one side of the reference level (end of the P R segment) is less than 0 5 mv in all of the bipolar extremity leads unipolar extremity leads and augmented unipolar extremity leads and less than 1 0 mv in all of the usually recorded *precordial leads* The term is to be used only when the criteria are met in all of the leads or in all those available

11 6 *Broad Q Wave* —The duration of the initial depression of the QRS group is 0 04 sec or greater The definition does not apply if the only QRS deflection is downward (QS)

11 7 *Elevation of S T Junction (J)* —The elevation of the junction compared to the reference level (end of the P R segment) exceeds 0 1 mv in the bipolar extremity leads or 0 05 mv in the unipolar extremity leads or 0 075 mv in the augmented unipolar extremity leads or 0 2 mv in the usually recorded *precordial leads*

11 8 *Depression of S T Junction (J)* —The depression of the junction compared to the reference level *(end of the P R segment)* exceeds 0 1 mv in the bipolar extremity leads or 0 05 mv in the unipolar extremity leads or 0 075 mv in the augmented unipolar extremity leads or 0 1 mv in the usually recorded precordial leads

11 9 *High Voltage of T Wave* —The voltage of the T wave measured in one direction from the reference level exceeds 0 7 mv *in* any of the bipolar extremity leads *or* 0 35 mv in any of the unipolar extremity leads or 0 5 mv in any of the augmented extremity leads or 2 0 mv in any of the precordial leads

TABLE IV*—NORMAL ADULTS 20 YEARS AND OVER SUPINE SIZE OF THE ELECTROCARDIOGRAPHIC DEFLECTIONS IN THE BIPOLAR EXTREMITY AUGMENTED UNIPOLAR EXTREMITY AND PRECORDIAL LEADS IS GIVEN IN TENTHS OF A MILLIVOLT

| Lead | P | | | | Q | | | | R | | | | S | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | No C | M | M | Mn | No C | M | Mx | Mn | No C | M | M | M | No C | M | M | M |
| I | 475 | 0 | 2 5 | 0 69 | 505 | 0 | 2 0 | 0 ?7 | 0 | 0 7 | 19 4 | 5 51 | 0 | 0 | 6 4 | 1 ?7 |
| II | 475 | 0 | 3 0 | 1 07 | 0 | 0 | 4 0 | 0 38 | 05 | 0 | 8 0 | 9 41 | 05 | 0 | 8 2 | 1 36 |
| III | 475 | —0 8 | 0 | 0 6 | 0 | 0 | 4 0 | 0 48 | 0 | 0 | 22 0 | 5 56 | 05 | 0 | 13 0 | 1 9 |
| $V_R$ | 3 | —1 0 | —0 5 | —0 63 | 6 | 0 | 8 0 | 48 | 6? | 0 | 3 0 | 0 90 | 6? | 0 | 11 0 | 3 01 |
| $V_L$ | 32 | —0 5 | 0 5 | 0 07 | 6? | 0 | 1 5 | 0 16 | 6 | 0 | 7 0 | 1 1 | 62 | 0 | 7 0 | .04 |
| $V_F$ | 3 | 0 | 0 | 0 ? | 62 | 0 | .0 | 0 30 | 6 | 0 | 1 0 | 6 8? | 6 | 0 | 6 5 | 0 74 |
| $aV_R$ | 411 | —1 5 | —0 1 | —0 79 | | 0 | 16 8 | 38 | | 0 | 4 1 | 0 94 | 5? | 0 | 1 7 | 3 76 |
| $aV_L$ | 411 | —1 0 | 1 4 | 0 51 | | 0 | 3 | 0 7 | | 0 | 10 1 | 2 61 | | 0 | 11 3 | 1 3 |
| $aV_F$ | 411 | —1 8 | 1 7 | 0 74 | | 0 | 0 | 0 38 | | 0 | 0 0 | 4 3 | 5 | 0 | 7 1 | 0 81 |
| $V_1$ | 371 | —1 1 | | 0 57 | 6 | 0 | 0 | 0 | 67 | 0 | 15 | 3 09 | 67 | 0 8 | 6 ? | 9 44 |
| $V_2$ | 3 1 | —0 7 | 0 | 0 60 | 94 | 0 | 0 | 0 | 94 | 0 | 3 0 | 5 96 | 94 | 0 | 9 | 14 09 |
| $V_3$ | 371 | —0 | 0 | 0 61 | 67 | 0 | 1 5 | 0 01 | 67 | 0 7 | 4 6 | 8 93 | 567 | 0 | ?7 5 | 9 51 |
| $V_4$ | 371 | —0 | 3 | 0 60 | 594 | 0 | 4 0 | 0 13 | 94 | 1 8 | 40 0 | 13 78 | 94 | 0 | 8 8 | 5 93 |
| $V_5$ | 371 | 0 | 4 | 0 6 | 67 | 0 | 3 4 | 0 43 | 67 | 0 4 | 33 6 | 1 01 | 67 | 0 | 16 1 | 1 96 |
| $V_6$ | 3 1 | 0 | 1 8 | 0 54 | 64 | 0 | ?7 | 0 44 | 64 | 0 | 6 | 9 68 | 64 | 0 | 14 3 | 1 00 |
| $V_E$ | — | — | — | — | 0 | 0 | 0 | 0 | 30 | 0 | 12 8 | 5 81 | 0 | 0 | 16 | 6 09 |

| Lead | RS or QR | | | | S—T | | | | T | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | No C | M | Mx | M | No C | M | M | M | No C | Mn | Mx | M |
| I | 63 | 3 0 | 0 6 | 8 54 | 100 | —0 3 | 0 9 | 0 11 | 505 | —0 5 | 5 6 | 0 |
| II | 63 | 8 0 | 3 0 | 15 14 | 100 | —1 0 | 1 0 | 0 ?1 | 505 | 0 | 8 0 | ?67 |
| III | 63 | 3 . | .0 | 10 6? | 100 | —0 6 | 0 8 | 0 04 | 505 | —?0 | 5 5 | 0 7 |
| $V_R$ | 6 | 3 5 | 1 0 | 6 0 | 3 | 0 | 0 | 0 | 6. | —4 0 | —0 5 | —1 65 |
| $V_L$ | 6 | 0 5 | 8 5 | 3 37 | 3? | 0 | 0 | 0 | 6 | —1 0 | 1 | 0 ?9 |
| $V_F$ | 6? | 3 5 | 16 | 7 77 | 3 | 0 | 0 | 0 | 62 | 0 | 4 6 | 1 40 |
| $aV_R$ | — | — | — | — | — | — | — | — | 479 | —5 5 | —0 | — 40 |
| $aV_L$ | — | — | — | — | — | — | — | — | 479 | —4 0 | 6 0 | 0 78 |
| $aV_F$ | — | — | — | — | — | — | — | — | 479 | —0 6 | 5 | 1 8 |
| $V_1$ | 63 | 6 6 | 3 0 | 14 99 | 33 | 0 | 0 5 | 0 01 | 4 | —4 0 | 1 | 0 84 |
| $V_2$ | 63 | 13 0 | 5 0 | 26 8 | 33 | 0 | 1 0 | 0 09 | 54 | — 6 | 18 0 | 4 70 |
| $V_3$ | 63 | 11 1 | 4 6 | 24 1 | 33 | 0 | 0 | 0 0 | 54 | — 0 | 1 0 | 5 16 |
| $V_4$ | 63 | 9 0 | 51 6 | 6 16 | 33 | 0 | 1 0 | 0 03 | 4 | —0 | 17 0 | 5 06 |
| $V_5$ | 63 | 10 0 | 36 4 | 19 31 | 33 | 0 | 0 | 0 | 54 | 0 | 11 0 | 3 83 |
| $V_6$ | .3 | 7 0 | 4 5 | 13 ?3 | 33 | 0 | 0 | 0 | 1 | 0 | 6 9 | 2 ?0 |
| $V_E$ | 30 | 5 6 | 4 | 11 91 | — | — | — | — | 30 | 0 | | 3 |

*The table is based on normal series studied by Kossmann and Johnston Kossmann and Goldberg Wilson and Nyboer Deeds and Barnes Vaquero Limon and Limon Myers Klein Stofer and Hiratzka Sokolow and Friedlander Katz e de Mello

• $V_E$ lead from tip of the ensiform cartilage

12 2 *QRS Form* —Abnormal in direction shape or area
12 3 *Junction of QRS Form and T Form* —Abnormally displaced
12 4 *T Form* —Abnormal in direction shape or area

13 MISCELLANEOUS
13 0 *Other Conditions Not Listed Above*

11 10 *Low Voltage of T Wave* —The voltage of the highest T wave measured in one direction from the reference level is less than 0 1 mv in all of the extremity leads and less than 0 2 mv in all of the usually recorded precordial leads The term is to be used only when the criteria are met in all of the leads or in all those available

11 11 *Unusual U Wave* —A prominent or inverted deflection immediately after the T wave is present in any of the usually recorded leads from the external surface of the body

11 12 *Electrical Alternans* —In a normal sinus or other supraventricular rhythm or tachycardia variations in the form of the ventricular complexes occur in alternate beats without variations in the duration of the QRS interval Most often the variation in form involves QRS and T (Fig 33) rarely T alone is affected

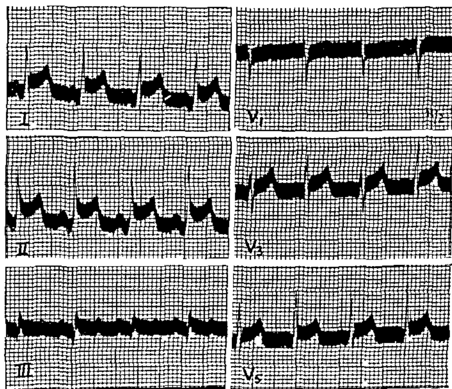



Fig 33

12 Spatial Vectorcardiogram*

12 1 *P Form* —Abnormal in direction shape or area

*Criteria for interpretation of the vectocardiogram are not yet available The method may be used with greater frequency in the future Therefore categories have been set aside in anticipation of this apparent trend even though criteria cannot be written

# NOMENCLATURE AND CRITERIA FOR THE PATHOLOGICAL DIAGNOSIS OF DISEASES OF THE HEART AND GREAT VESSELS



# NOMENCLATURE FOR PATHOLOGICAL DIAGNOSIS

## CONGENITAL ANOMALIES OF THE HEART AND GREAT VESSELS

ANOMALIES OF THE HEART AS A WHOLE

1 Dextrocardia

2 Dextroposition

ANOMALIES OF THE INTERATRIAL SEPTUM

3 Patent foramen ovale (persistent ostium secundum)

4 Patent ostium primum (ostium atrioventriculare)

5 Absence of interatrial septum total or subtotal (cor triloculare biventriculare)

6 Anatomic complications of interatrial septal defects

ANOMALIES OF THE INTERVENTRICULAR SEPTUM

7 Congenital aneurysm of the interventricular septum

8 Defect of the interventricular septum (Maladie de Roger)

9 Absence of interventricular septum total or subtotal (cor triloculare biatriatum)

ANOMALIES OF THE CHAMBERS OF THE HEART

10 Hypoplasia of the left heart chambers

11 Hypoplasia of the right heart chambers

ANOMALIES OF CORONARY ARTERIES

12 Origin of the left coronary from the pulmonary artery

13 Coronary arteries in transposition

14 Variation in distribution of coronary arteries

ANOMALIES OF THE MYOCARDIUM

15 Primary congenital rhabdomyoma of the heart

16 Congenital hypertrophy of the heart

ANOMALIES OF THE CONDUCTION SYSTEM

17 Congenital heart block partial or complete

ANOMALIES OF ENDOCARDIUM AND VALVES

18 Congenital bicuspid aortic valve

19 Stenosis or atresia of the pulmonary valve

20 Subpulmonary stenosis (Infundibular stenosis)

21 Subaortic stenosis

22 Aortic stenosis



51 Arteriosclerosis of the coronary arteries
52 Aneurysms of the coronary arteries
53 Embolism of the coronary arteries
54 Rupture (spontaneous) of the coronary arteries

## DISEASES OF THE MYOCARDIUM*

56 Abscesses of the myocardium
55 Acute myocarditis
83 Air embolism of heart
66 Amyloid infiltration of the heart
69 Aneurysmal dilatation of the heart *(aneurysm of the heart)*
58 Atrophy of the heart
59 Brown atrophy
64 Calcification of the myocardium
73 Dilatation of the myocardium
71 Enlargement of the heart
82 Fat embolism of myocardium
60 Fat infiltration of the myocardium
61 Fatty degeneration of the myocardium
67 Fibrosis of myocardium
65 Glycogenosis (von Gierke's disease)
74 Granulomata of the myocardium infections
76 Hemochromatosis of the myocardium
72 Hypertrophy of the myocardium
77 Hyperthyroidism The heart in
57 Idiopathic myocarditis (Fiedler's myocarditis)
68 Infarct of the myocardium
81 Injury of the myocardium
63 Myocardial degeneration in Friedreich's ataxia
78 Myxedema The heart in
80 Neoplasms of the myocardium (indicate neoplasm)
75 Parasitic lesions of the myocardium
84 *Poisoning of the heart (indicate specific agent if possible)*
70 Rupture of the myocardium spontaneous
62 Toxic myocardial degeneration *(indicate etiological agent if possible)*
79 Vitamin B deficiency The heart in

* The titles dealt with in this chapter have been arranged alphabetically in the nomenclature but are numbered according to their sequence in the text.

23 Aortic atresia
24 Tricuspid insufficiency
25 Endocardial fibro elastosis (Fetal endomyocarditis)

Anomalies of the Aortic and Pulmonic Trunks
26 Transposition of arterial trunks
27 Persistent truncus arteriosus communis partial or complete

Anomalies of the Ductus Arteriosus
28 Patent ductus arteriosus

Anomalies of the Aortic Arch
29 Retro esophageal right subclavian artery
30 Right aortic arch
31 Double aortic arch
32 Coarctation of the aorta adult type
33 *Coarctation of the aorta infantile type*

Anomalies of the Veins
34 Persistent left superior vena cava
35 Anomalies of the pulmonary veins

## RHEUMATIC HEART DISEASE

36 Rheumatic myocarditis
37 Rheumatic valvulitis and endocarditis
38 Rheumatic endocarditis of the atrium
39 Rheumatic pericarditis
40 *Rheumatic aortitis and pulmonary arteritis*
41 Rheumatic arteritis of the smaller arteries

## SYPHILIS OF THE HEART AND AORTA

42 Syphilitic aortitis
43 Syphilitic aneurysm of the aorta
44 Syphilitic involvement of the aortic valve
45 Coronary artery involvement in syphilitic aortitis
46 *Myocardium in syphilis*
47 Congenital syphilis

## DISEASES OF THE CORONARY ARTERIES

48 Arteritis of the coronary arteries (*endarteritis mesarteritis peri arteritis*)
49 Panarteritis nodosa (polyarteritis nodosa periarteritis nodosa)
50 Thromboangiitis obliterans of the coronary arteries

## DISEASES OF THE PERICARDIUM

115 Pericarditis
116 Adherent pericardium
117 Hemopericardium
118 Hydropericardium
119 Adipose changes in the pericardium
120 Uremic pericarditis
121 Pericarditis following myocardial infarction
122 Acute non specific pericarditis
123 Rheumatic pericarditis
124 Bacterial infection of pericardium
125 Tuberculous pericarditis
126 Fungus infection of pericardium
127 Pericardial injuries
128 Primary neoplasms of pericardium

## DISEASES OF THE CONDUCTION SYSTEM

85 Degenerative changes
86 Inflammatory lesions
87 Congenital lesions
88 Neoplasms

## DISEASES OF THE ENDOCARDIUM AND VALVES

89 Acute bacterial endocarditis
90 Subacute bacterial endocarditis
91 Indeterminate endocarditis
  a Atypical verrucous endocarditis (Libman Sacks)
  b Thromboendocarditis
92 Tuberculous endocarditis
93 Syphilitic endocarditis
94 Annular sclerosis
95 Valvular (endocardial) atheroma
96 Valvular sclerosis
97 Mural endocardial fibrosis
98 Rheumatoid valvulitis
99 *Mural thrombosis*
100 Endocardial blood cysts
101 Anatomical signs of valvular disease
  a Insufficiency
  b Stenosis of the orifice of a valve
  c Endocardial pockets

## DISEASES OF THE AORTA AND PULMONARY ARTERY

102 Acute aortitis
103 Giant cell mesaortitis
104 Arteriosclerosis of the aorta or pulmonary artery
105 Calcification of the aorta
106 Medial necrosis of the aorta
107 Aneurysm of the aorta or pulmonary artery
108 Rupture of the aorta (spontaneous)
109 Dissecting aneurysm
110 Thrombosis of the aorta
111 Embolism of the aorta
112 Thrombosis and embolism of the pulmonary artery
113 Injury of the aorta
114 Arteriovenous fistula

CHAPTER 1

# CONGENITAL ANOMALIES OF THE HEART AND GREAT VESSELS

Owing to lack of standardization in terminology adequate figures of relative incidence of the specific varieties of anomaly are not available According to data covering some three hundred consecutive autopsies the major groups (Abbott classification) have the following frequency

I Without cyanosis 24%
II With late cyanosis 44%
III Cyanosis from birth 32%

The most common major anomalies encountered at necropsy are

1 Anomalies of interatrial septum
2 Anomalies of interventricular septum
3 Coarctation of aorta adult type
4 Fallot or Eisenmenger tetralogies
5 Transposition of arterial trunks

These categories include about nine tenths of all cases Application of more rigid diagnostic criteria might reduce the incidence of patent foramen ovale as an autopsy finding

## AIMS AND PROCEDURES IN NECROPSY STUDY OF CONGENITAL ANOMALIES OF THE HEART

Evaluation of a congenital anomaly of the heart by the pathologist should include not only a morphological study but also an attempt at reconstruction of the altered circulation and its possible effect on the production of clinical signs and symptoms Thus a venous arterial shunt with resultant clinical cyanosis may be found dependent upon an anatomical defect (e g a defect of the interatrial septum) plus a functional alteration in pressure such as an increase of pressure in the right atrium from failure causing flow from right to left. Care must be exercised in correlation and reconstructions of function must be regarded as only putative

Reconstruction of the embryologic aberration or pathogenesis of the anomaly in question is another prerogative of the pathologist and again this should be with realization of its speculative nature

Helpful special methods of examination include

be diagnostic for the structurally left ventricle though the bundle branches are seldom distinctly visible The number of cusps in mitral and tricuspid valves is *not* a valid criterion of laterality as variation in position of the atrioventricular orifices influences the form of their cusps

2 DEXTROPOSITION —Displacement of the heart toward the right or even into the right hemithorax may occur with congenital hernia or defect of the diaphragm The heart is neither inverted nor malformed

## ANOMALIES OF THE INTERATRIAL SEPTUM

The ostium primum a residuum of the original communication between the two atria is a defect in a lower part of the septum primum bordering the original communication between the two ventricles The entire space connecting atria and ventricles is the ostium atrioventriculare commune The term is practically synonymous with ostium primum because defects of the lower interatrial septum almost invariably involve the margin of the interventricular septum and atrioventricular valves In like manner the terms foramen ovale and ostium secundum are synonyms in common use despite precise embryological distinction Both terms cover defects in the upper part of the interatrial septum

3 PATENT FORAMEN OVALE (persistent ostium secundum) —Probe patency is found in a fourth of all adult hearts yet functional closure is maintained by the valve of the foramen ovale Normally functional closure begins at birth although anatomic closure continuing to obliteration does not reach its maximum until near the end of the first postnatal month or even the last third of the first postnatal year In description of specimens distinction should be made between anatomic or probe patency with adequate valve and presumptive functional patency without adequate valve Only the latter may be considered significant as a congenital anomaly

Anatomical or functional patency of the foramen ovale may coexist with persistent ostium primum as a double septal defect More frequently however the foramen ovale itself may be divided into two or more apertures separated by filamentous or membranous bands owing either to overgrowth of the septum secundum or to too great resorption of the septum primum The criteria of persistent ostium primum (see below) should be rigidly applied in the differential diagnosis of multiple defects of interatrial septum Large defects of the foramen ovale occur with some frequency in cases of arachnodactylism (Marfan s syndrome)

1 Routine removal of the heart lungs and thoracic aorta from the body in block in cases where a congenital anomaly of the heart and great vessels is suspected to preserve relationships

2 *Perfusion of the heart chambers with fluid via each atrium in turn* the aortic and pulmonary trunks having been previously incised to check relative volume flow through the chambers and septal defects if present

3 Sectioning the heart in multiple planes paralleling that of the valve orifices and perpendicular to that of the long axis of the heart Cuts of this type may be alternated from anterior and posterior surfaces and if carried only partially through the walls opposite those of entry will allow the heart to be opened for study then folded back again accordion fashion for storage This method of sectioning displays *relative size and position of septa septal defects and cavities* without the distortion attendant on ordinary methods of opening the heart

4 Sectioning the heart in a coronal plane best demonstrates the overriding character of the aorta in relation to a subaortic septal defect

5 Cast impressions of heart cavities are sometimes useful

6 *Injection corrosion preparations of the pulmonary vascular tree* are of interest in cases of Fallot or Eisenmenger tetralogies

## ANOMALIES OF THE HEART AS A WHOLE

1 Dextrocardia —Mirror image inversion of the heart and great vessels occurs *with* complete situs inversus viscerum

*Inversion of the heart without accompanying inversion of* other viscera is usually combined with other malformations of the heart especially one of the types of transposition of aortic and pulmonary trunks Isolated inversion of the heart and varying degrees of inversion of other viscera are frequent in the rare cases of corrected transposition

The diagnosis of structural inversion of the heart or of one of its segments depends *upon the finding of inverse laterality of one or more* of the key structures In the atrial segment the topography of the septum is diagnostic the valve of the foramen ovale normally marking the left side with the crista terminalis or limbus fossae ovalis on the right side In the ventricular segment the critical landmark is the crista supraventricularis a muscle ridge normally present in the roof of the pulmonary conus The Y shaped left bundle branch lying in the wall of the interventricular *septum just beneath the endocardium* may

Displacement of the aortic orifice to the right—that is dextroposition of the aorta—may be relative rather than real This type of malformation is an integral part of the tetralogies of Fallot or Eisenmenger

9 ABSENCE OF INTERVENTRICULAR SEPTUM TOTAL OR SUBTOTAL (cor triloculare biatriatum or cor triloculare monoventriculare) is an infrequent malformation occurring usually in combination with either transposition of arterial trunks or persistent truncus arteriosus communis In suspected cases careful search of the specimen must be made to exclude the presence of a small second ventricle embedded or concealed in the wall of the main (and apparently solitary) ventricle Sectioning the heart in planes perpendicular to the ventricular axis may be helpful

## ANOMALIES OF THE CHAMBERS OF THE HEART

10 HYPOPLASIA OF THE LEFT HEART CHAMBERS —In a characteristic but uncommon malformation the left ventricle is rudimentary The mitral and aortic orifices are minute or atretic The ascending aorta is so small that it may escape notice unless the coronary ostia are sought The entire complex may be regarded as a result of *primary hypoplasia* of the left side of the bulboventricular loop of the embryonic heart

11 HYPOPLASIA OF THE RIGHT HEART CHAMBERS —In a relatively rare anomaly forming an approximate counterpart of the *combination* just described the right ventricle is rudimentary with marked stenosis or atresia of tricuspid and pulmonary orifices An interatrial septal defect is the rule in hypoplasia of either the left or the right chambers of the heart

## ANOMALIES OF CORONARY ARTERIES

Considerable variation occurs in the position of the ostia of both coronary arteries in relation to the sinuses of Valsalva

12 ORIGIN OF THE LEFT CORONARY FROM THE PULMONARY ARTERY —Owing to insufficient supply of oxygenated blood in postnatal life *the myocardium* of the left ventricle may show *necrosis* perhaps with deposition of calcium a notable *occurrence in infancy* Dilatation of the left ventricle and localized softening of the myocardium are characteristic features in the *gross* The basic anomaly may escape notice unless specifically sought

4 Patent Ostium Primum (ostium atrioventriculare) —This is a rounded or semilunar defect usually of fair size situated in the lowest portion of the interatrial septum extending part way into the posterior part of the interventricular septum There is usually cleavage of the anterior (septal) cusp of the mitral valve into two segments and fusion of adjoining leaflets of the tricuspid and mitral valves through the septal defect Probes can be passed obliquely through the defect from each atrium into the contralateral ventricle Septal defects of this type are one of many features in the syndrome of Mongolian idiocy but many occur without this association

5 Absence of Interatrial Septum Total or Subtotal (cor triloculare monatriatum or cor triloculare biventriculare) is infrequent

6 Anatomic Complications of Interatrial Septal Defects — Sizeable functional defects of the upper interatrial septum are usually *accompanied by dilatation of the right atrium and ventricle and of the* pulmonary artery with relative narrowing of the aorta

The combination of mitral stenosis probably rheumatic with inter*atrial septal defect is commonly designated Lutembacher s syndrome*

## ANOMALIES OF THE INTERVENTRICULAR SEPTUM

7 Congenital Aneurysm of the Interventricular Septum is an evagination of the membranous portion of the septum into the right ventricle or right atrium Gross and microscopic evidence of inflammation must be eliminated in distinguishing a congenital aneurysm of the septum from a mycotic aneurysm

8 Defect of the Interventricular Septum (Maladie de Roger) — A localized or small defect of the muscular septum is usually situated near the base of the heart just anterior to the membranous portion of the septum Some of the smallest openings in this position may be functionally closed by the adjoining septal cusp of the tricuspid valve

This anomaly constitutes the maladie de Roger It permits an arteriovenous shunt of *blood into the right ventricle A patch of endo*cardial thickening in the wall of the right ventricle opposite the septal defect is attributed to the stimulus of the impact of the stream of shunted *blood impinging on it*

Other defects of the membranous portion of the interventricular septum are located beneath the aortic orifice so that the orifice appears *to override the* defect with free access into it from both ventricles

## ANOMALIES OF THE CONDUCTION SYSTEM

17 CONGENITAL HEART BLOCK PARTIAL OR COMPLETE may appear in defects of the interventricular septum With such defects the bundle of His may be interrupted developmentally though it is usually preserved lying in the posterior wall of the septal defect In other cases the anatomical basis of the block may not be demonstrable (see Chapter VI)

## ANOMALIES OF THE ENDOCARDIUM AND VALVES

18 CONGENITAL BICUSPID AORTIC VALVE.—The two cusps may be well formed and identifiable as a congenital anomaly There may be all grades of incomplete fusion between the component halves of one of the two cusps so that it is difficult to distinguish between instances of congenital fusion and cases of fusion due to acquired disease rheumatic valvulitis or bacterial endocarditis both diseases to which the congenital anomaly predisposes

The differentiation of the acquired and congenital forms depends upon the nature of the commissural ridge The congenital ridge is a sharply outlined hemicylindric elevation of the aorta in the sinus of Valsalva with parallel lateral borders and a smoothly rounded surface descending almost vertically from the upper limit of the sinus to its floor The acquired ridge on the contrary presents lateral surfaces that diverge in the direction of the free margin of the cusp has a free surface that may be fissured longitudinally and descends obliquely downward in the sinus of Valsalva within which it is only moderately depressed Bicuspid valves in adults are commonly thickened calcified and stenotic

Microscopically vertical sections taken in a plane perpendicular to that of the aortic wall show the congenital ridge to contain elastic lamellae derived from the aorta with a whorling configuration in the center of the ridge The elastica has an abnormally low insertion into the annulus fibrosus and may terminate superficial to it In the ridge derived from inflammatory fusion the usual commissural relationships are preserved in that the annulus is superficial to the elastica

19 STENOSIS OR ATRESIA OF THE PULMONARY VALVE —Uncomplicated stenosis of the pulmonary valve is rare Atresia of the pulmonary valve without accompanying defect of the interventricular septum is a part of the syndrome of hypoplasia of the right chambers of the heart

Stenosis or atresia of the pulmonary valve accompanied by a defect

13. CORONARY ARTERIES IN TRANSPOSITION.—In crossed transposition of arterial trunks there is inversion of the situation in the normal heart. The left coronary artery arises from the sinus of Valsalva situated to the right, while the right coronary arises from the left sinus of Valsalva.

14. VARIATION IN DISTRIBUTION OF CORONARY ARTERIES.—Normal variation in the distribution of the coronary arteries concerns principally the relative preponderance of the right coronary and the posterior circumflex branch of the left coronary in supplying the musculature of the posterior wall of the left ventricle. There is similar variation in the extent of the anterior descending branch of the left coronary over the apex. Accessory right coronary arteries are frequent. Special methods of injecting the coronary arteries and special procedures for opening the heart should be employed for the study of coronary arterial distribution (see Chapter IV).

## ANOMALIES OF THE MYOCARDIUM

15. PRIMARY CONGENITAL RHABDOMYOMA OF THE HEART is a tissue malformation (hamartoma) rather than a true neoplasm. Metastasis or even invasion has never been observed. One or more nodules of *yellow white firm tissue protrude beneath the endocardial surface* of the ventricles or atria.

Microscopically these nodules consist of abnormal cardiac muscle with prominent spider cells, which are giant rounded cells containing myofibrillae radiating from the centrally placed nucleus. Between the myofibrillae are large vacuoles containing glycogen. The myofibrillae may appear in ordinary hematoxylin-eosin sections but are seen to better advantage with phosphotungstic acid hematoxylin. Tuberous sclerosis of the brain is a common finding in cases of rhabdomyoma of the heart.

16. CONGENITAL HYPERTROPHY OF THE HEART.—In von Gierke's disease the heart is uniformly enlarged and contains greatly increased amounts of glycogen of a form that is more stable than usual so that it may sometimes be stained even after formalin fixation. Increased amounts of glycogen are present in the liver and kidneys. In many cases there is also much lipid within the liver, and the fresh blood may have a milky appearance (*lipemia*).

cusps are generally thickened and bulbous resembling those in the embryo before the definitive stage of cusp formation is reached Thickening of the endocardium of the type to be described under endocardial fibro-elastosis is frequently a concomitant finding

23 Aortic Atresia —Atresia of the aortic valve is a part of the syndrome of hypoplasia of the left heart described above

24 Tricuspid Insufficiency —Ebstein's disease is a structural disturbance of the entire right heart The tricuspid valve is displaced obliquely downward into the right ventricle the line of attachment of the valve cusps being roughly vertical instead of transverse The anterior cusp retains its attachment to the annulus fibrosus while the posterior cusp is attached instead to the walls of the right ventricle The posterior and septal cusps are shortened and bound down with underdevelopment of the corresponding papillary muscles The large part of the right ventricle that lies above the valve is functionally part of the right atrium This part of the ventricle along with the anatomic right atrium is dilated owing to incompetency of the malformed valve The syndrome is rare but characteristic.

25 Endocardial Fibro-Elastosis (endocardial hyperplasia endocardial sclerosis fetal endocarditis or fetal endomyocarditis) —Lesions of this group are probably more frequent than present knowledge indicates because of classification under other headings The endocardium is thickened and more resistant than usual It is opaque white or cream colored The area affected may include only the left surface of the muscular interventricular septum toward the base of the heart or the entire inner surface of the left ventricle may be involved The endocardium of the left atrium is next most frequently affected The cusps of the aortic and mitral valves may be thickened with shortening and thickening of the chordae tendineae The left ventricle is usually dilated with some hypertrophy With aortic atresia the left ventricle is hypoplastic

Microscopically the thickened endocardium shows great increase in fibro elastic tissue The myocardium shows a variety of changes including degeneration of muscle cells fibrosis and deposition of particulate calcium The classical theory of pathogenesis is that the lesions represent the end stage of a healed antenatal inflammatory process According to the newer alternative explanation the fibro-elastosis of the endocardium and valves is a tissue malformation or primary hyperplasia which mechanically blocks the orifices of the luminal vessels

of the interventricular septum with dextroposition of the superjacent aortic orifice and with hypertrophy of the musculature of the right ventricle comprises the relatively common *tetralogy of Fallot* There may be a reduction in the number of cusps of the pulmonary valve About a fifth of the cases of this complex present an additional feature the arch of the aorta courses over the right main bronchus instead of the left with inverse order of the origin of its branches The presence or absence of this added malformation is of considerable importance in operative procedures for correction of pulmonary stenosis

It is important to distinguish from the tetralogy of Fallot the anatomically similar but functionally dissimilar *tetralogy of Eisenmenger* Here though there is a defect of the interventricular septum with overriding aortic orifice and hypertrophy of the right ventricle the pulmonary valve is of normal size or larger and the pulmonary artery and its branches showing striking dilatation

Microscopically multiple thromboses of the smaller branches of the pulmonary arterial tree within the lung have been reported as characteristic of the tetralogy of Fallot In sections of lung from cases of the Eisenmenger tetralogy dilatation of the pulmonary arterial branches may be of diagnostic value

20 Subpulmonary Stenosis (conus stenosis infundibular stenosis) —In this condition the stenosis affects the lower portion of the pulmonary conus A band of muscle with overlying fibrous thickening of the endocardium encircles the right ventricle in the area indicated The portion of the cavity of the right ventricle lying between this stenosis and the pulmonary orifice may be expanded into a small accessory chamber The anomaly may occur as part of the tetralogy of Fallot with or without an accompanying valvular pulmonary stenosis

21 Subaortic Stenosis —This rare malformation is homologous to the subpulmonary stenosis just described but the stenosis is not as marked nor is the distance from valve to stenosis as great The part of the left ventricle situated just below the aortic orifice is narrowed and encircled by a fibrous ridge that projects into the lumen The ridge may cross completely or incompletely the aortic or anterior cusp of the mitral valve

22 Aortic Stenosis —The orifice of the usual type of bicuspid aortic valve is characteristically of normal caliber In stenosis of the aortic orifice with or without reduction in the number of cusps the

The pathologic study of any heart manifesting changes of transposition should include description of the interior topography of ventricles and atria with reference to the criteria of inversions stated (under Dextrocardia) description of the coronary arterial system and notation of possible structural inversions elsewhere in the body

27 PERSISTENT TRUNCUS ARTERIOSUS COMMUNIS partial or complete—The aortic and pulmonic trunks are combined in a common trunk with or without partial delimitation of the component channels by a rudimentary septum In diagnosis the following criteria are to be observed

a Only one large arterial trunk must be present Careful search is sometimes necessary to exclude presence of a rudimentary aortic trunk extending along an apparent common trunk

b The common trunk must give rise to the coronary arteries and to the pulmonary arteries (except in the absence of pulmonary arteries)

c A defect of the interventricular septum is regularly present

d The common trunk arises from a site above the septal defect or arises solely from the right ventricle

e The orifice of the common trunk should have four cusps with the coronary ostia situated above opposite cusps Although this is not always found it is an important criterion when present

f The crista supraventricularis stops at the base of the septum beneath the orifice of the common trunk

g One of the forms of interatrial septal defect is generally present

Two other types of anomalies are subject to confusion with persistent truncus communis In aortic atresia or hypoplasia of the left cardiac chambers the aortic trunk is reduced to a rudimentary easily overlooked vessel supplying the coronary arteries in retrograde fashion from the ductus arch of the aorta or innominate artery The term solitary pulmonary trunk has been applied to the remaining large arterial vessel in these instances

In pulmonary atresia or in hypoplasia of the right cardiac chambers the pulmonary arteries may be absent or may arise from the aortic trunk The term solitary aortic trunk has been applied to the remaining large arterial vessel in such cases

## ANOMALIES OF THE DUCTUS ARTERIOSUS

28 PATENT DUCTUS ARTERIOSUS—As in the case of the foramen ovale normal anatomic closure of the ductus arteriosus follows func

this in turn causes dilatation of the intramyocardial capillaries producing stagnation anoxemia and degenerative changes within the myocardium

## ANOMALIES OF THE AORTIC AND PULMONIC TRUNKS

26 Transposition of Arterial Trunks—In transposition either *the aorta alone or the* aortic and pulmonary trunks arise from incorrect ventricles The relationships of aortic and pulmonary trunks are thus transposed Four main types of transposition are designated in the classification of Spitzer

Type I Overriding aorta or tetralogy of Fallot—Criteria as given above under Stenosis or atresia of the pulmonary valve

Type II Simple transposition of aorta—The aortic and pulmonary trunks both arise from the right ventricle There is usually a large defect in the interventricular septum and stenosis of the pulmonary valve

Type III Crossed transposition of aorta—The aorta arises from the right ventricle the pulmonary artery from the left The aortic orifice lies in front of the pulmonary orifice The interventricular septum may be intact or almost entirely deficient (cor triloculare biatriatum)

Type IV *Mixed transposition of aorta*—The aorta *arises from the* right ventricle the pulmonary artery from the left The aortic orifice lies in front of the pulmonary orifice The aorta may be stenotic There *is an interventricular septal defect Mitral* and *tricuspid orifices both* open into the left ventricle

Gradations occur between these types A particular pattern intergrading between transposition Types II and III characterized by transposition of the aorta to the right ventricle with origin of the pulmonary artery in overriding position above a defect of the interventricular septum has received special recognition as the *Taussig Bing heart*

There are also instances of corrected transposition (Types II III and IV) in which although the arterial trunks arise from the correct ventricles transposition is evident in the disturbed relationship of these vessels the aorta arising in front of the pulmonary artery instead of behind it The correction may be anatomic in that the aorta joins the left ventricle and the pulmonary artery joins the right ventricle The correction may be functional " in that the aorta receives blood coming from pulmonary veins while the pulmonary artery receives blood from the systemic veins owing to inversion of the atria and great veins (see under Dextrocardia )

Finally 15 per cent of cases of right aortic arch occur in conjunction with a left aortic arch one of the forms of double aortic arch considered in the next section

31 Double Aortic Arch—An aortic arch courses over each bronchus fusing posteriorly into a single descending aorta In most of the cases of this rare anomaly the right arch is distinctly larger than the left A double aortic arch constitutes a ring completely encircling the trachea and esophagus A similar situation exists in cases of right aortic arch without inversion of other viscera with or without retro-esophageal left subclavian artery for the left sided ductus arteriosus in effect completes the ring encompassing the trachea and esophagus

Pathologic description of a case of aortic arch anomaly should include specific mention of all of the branches of the arch of the ductus arteriosus and of the situs of the other viscera

32 Coarctation of the Aorta Adult Type—In this condition there is fairly abrupt narrowing ranging to complete occlusion of the descending aorta in the region of the usually obliterated ductus arteriosus with resultant formation of an extensive collateral circulation between the branches of the aorta arising proximal to the constriction and the branches of the aorta arising distally The intercostal arteries show tortuosity and dilatation that is most marked toward their posterior ends Erosion or notching of the inferior margins of adjoining posterior portions of the ribs is characteristic The aortic valve is frequently bicuspid There is usually hypertension in the portion of the aorta proximal to the coarctation with resultant dilatation of the ascending aorta and arch Multiple miliary or berry type aneurysms may occur in the junctional angles of the circle of Willis or of the cerebral arteries

In microscopic sections of portions of aorta removed in the operative correction of coarctation by excision and direct re anastomosis arteriosclerotic changes are prominent above the coarctation and relatively scant below the level At the constriction there is deficiency of elastica and muscle with increase in fibrous tissue

A distinct preponderance of males is observed in cases of coarctation of the aorta (adult type) in contra-distinction to an equal preponderance of females in cases of uncomplicated patency of the ductus arteriosus

tional closure The obliterative process does not reach its maximum *until the second postnatal month* In three fourths of all cases the ductus is closed at the end of the first trimester of the first postnatal year and in over 95 per cent by the end of the first year This anatomic closure is *effected by hypertrophy and hyperplasia of the intima* of the ductus with collagen tissue infiltration Thrombosis may provide a subsidiary mechanism While strict limitation of size would provide only an arbitrary criterion for diagnosis patency of the ductus can scarcely be considered of pathologic significance in infancy and is entirely meaningless in the neonatal period Distinction should be made (as in foramen ovale) between minute anatomic patency and broad (presumptive functional if past early infancy)) patency In the latter the internal diameter may range from 1 to 10 millimeters Here again quantitative criteria would be only arbitrary

About two thirds of the cases of *abnormally patent* ductus arteriosus are complicated by the presence of some other cardiac anomaly In about a third of the *uncomplicated* cases bacterial endocarditis supervenes characteristically best developed at the pulmonary end of the ductus

## ANOMALIES OF THE AORTIC ARCH

29 Retro Esophageal Right Subclavian Artery —The right subclavian artery arises as the last branch of the aortic arch and passes behind the esophagus and trachea to reach the right side of the thoracic outlet

30 Right Aortic Arch —In about a half of the cases of this anomaly there is also a general visceral inversion (situs inversus viscerum totalis) The aortic arch and its branches present a mirror picture of the normal

In another 20 per cent of the cases of right aortic arch the vessel and its branches show mirror image inversion without corresponding inversion of other viscera Many of these cases are associated with intracardiac anomalies especially with the tetralogy of Fallot

In a third group of cases (25 per cent) the aortic arch courses over the right bronchus the left subclavian artery arises as *the last branch* and passes behind the trachea and esophagus to reach the left side of the thoracic outlet This type of anomaly essentially a mirror picture of the more *common retro esophageal right subclavian artery* is generally not associated with intracardiac malformation

## CHAPTER II

# RHEUMATIC HEART DISEASE

IT HAS BEEN clearly demonstrated that rheumatic fever involves not only the heart but the vessels including the capillaries Furthermore lesions that are quite similar to those in the heart are found in the vascular tree

The agent producing rheumatic fever has not been definitely established. However the occurrence of a specific type of lesion in patients presenting the typical signs and symptoms of the disease has permitted the diagnosis of this disease by the identification of these lesions

Many of the terms so useful clinically in denoting the duration of the disease are of little value to the pathologist Lesions may occur without causing any clinical evidence of the disease or the disease may be slowly progressive In the presence of acute lesions there are often found healed healing and acute lesions adjacent to each other In some instances only completely healed lesions are seen the location and shape of these scars are sufficiently characteristic to enable one to diagnose them as resulting from rheumatic fever When only the scars are found the lesion should be designated as healed



Fig 1 Rheumatic myocarditis Early Aschoff nodule In the center is swollen fragmented collagenous tissue about the periphery are small mononuclear cells and a few Aschoff cells

33 Coarctation of the Aorta Infantile Type —The narrowing is diffuse and involves the aortic isthmus above the point of insertion of the *usually patent* ductus arteriosus This type of coarctation is less common than the adult type and customarily is combined with intra cardiac malformation

Intergrades between the adult and infantile forms of coarctation occur, so that the distinction is often arbitrary In all cases of coarcta tion patency or non patency of the ductus should be determined be cause of its important functional significance

## ANOMALIES OF THE VEINS

34 Persistent Left Superior Vena Cava —This malformation rep resents simple persistence of a lumen in the fetal vessel that normally closes to form the ligament of Marshall The left superior cava drains into the right atrium *via* the coronary sinus

35 Anomalies of the Pulmonary Veins —One or more of the pul monary veins may drain into the right atrium instead of into the left Descriptions of cases should specify the topography of the interatrial septum as this is of consequence in considerations of pathogenesis of the anomaly

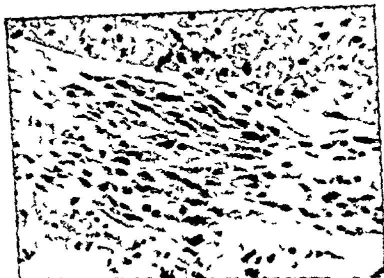


FIG 3 Rheumatic myocarditis Healing Aschoff nodule The Aschoff cells have become spindle shaped and resemble connective tissue cells



FIG 4 Acute rheumatic endocarditis Mitral valve A continuous chain of verrucae along line of closure

36 Rheumatic Myocarditis—The characteristic lesion in the myocardium is the Aschoff nodule This submiliary nodule is the most specific lesion of rheumatic fever

Early changes are swelling and fragmentation of collagen bundles Small mononuclear cells collect about these fragmented bundles of collagen with these cells there may be a few polymorphonuclear leucocytes (Fig 1) Later there appear the large mononuclear cells that have a faintly basophilic cytoplasm and a large vesicular nucleus with a prominent chromatin mass Some of these cells are multinucleated The prominent large cells are the so called Aschoff cells as they collect the smaller cellular elements disappear and the well developed nodule consists only of the Aschoff cells The cells of the nodule spread apart the reticulum fibers (Fig 2)

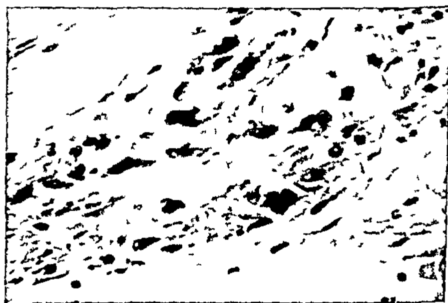


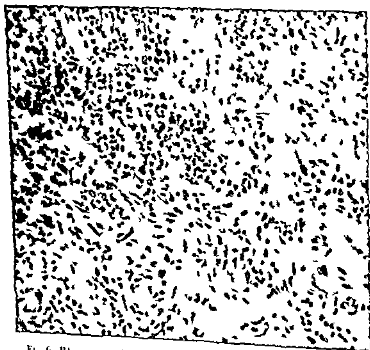
Fig 2 Rheumatic myocarditis Aschoff nodule Remnants of swollen fragmented collagen in center with Aschoff cells

As the nodule becomes older the characteristic cells are more spindle shaped and resemble more closely connective tissue cells (Fig 3) When the nodule is completely healed there remains a dense vascular scar

Aschoff nodules are found in the myocardium close to branches of the coronary arteries especially in the posterior portion of the left ventricle and in the interventricular septum they also are seen in the valves endocardium pericardial tissues and the atrioventricular conduction system (Fig 40)

and mitral valves less frequently on the tricuspid valve and occasionally on the pulmonary valve Sometimes on the mitral valve the vegetations are broader and flatter and extend across the atrial surface of the leaflets for a distance of 5 or 6 millimeters Vegetations are rarely found on the ventricular surface of the atrioventricular valves The vegetation is composed of granular material staining deeply with eosin (Fig 5) In the earliest vegetations this material extends down into the underlying valve substance for a short distance Fibrin can be demonstrated at the surface where the vegetation is in contact with the blood and at the base where it joins the valve substance the fibrin is however only in small amounts

At the base of the vegetation are collected large mononuclear cells Fibroblasts from the underlying valve penetrate into the eosinophilic material composing the vegetation Sometimes small compact masses of this material lie embedded in this newly formed connective tissue with more recent deposits of the granular material on the surface Ultimately the granular material is completely replaced by connective



Fi 6 Rheumat c valvul t s The su bstance of the leaflet is heavily infiltrated by polymo phonuclear leucocytes and lymphocytes Fibrol la s an l cap illaries are increased in the leaflet.

Recent and healed nodules are present in many hearts indicating more than one attack of myocarditis or a long continued period of inflammation Unhealed nodules are often found in rheumatic cases when there has not been any clinical evidence of recurrence or recent activity of the disease They also may be discovered in the hearts of individuals who have never had rheumatic polyarthritis and who died of some other disease

The nodules are not confined to the heart They have been found in the adventitia of the pulmonary artery and aorta the diaphragm periesophageal tissues and galea aponeurotica also collections of cells resembling the Aschoff nodule have been described in the lungs in the peritoneum and near the joints

37 Rheumatic Valvulitis and Endocarditis —The term *rheumatic valvulitis* may be used to denote the changes that occur in the substance of the valve and the vegetations that form upon it

The vegetations are small firm and wartlike the recently formed verrucae are dull yellowish and the surface is rough They are firmly adherent to the valve and do not break away hence infarcts in distant organs are not found in uncomplicated rheumatic heart disease

The vegetations often form a continuous chain along the line of closure of the valves (Fig 4) occurring most commonly on the aortic



Fig 5 Rheumatic endocarditis Mitral valve A healing vegetation composed of eosin staining material with ingrowth of connective tissue at base



Fig Mitral stenosis due to rheumatic endocarditis The valve orifice is greatly reduced in size Chordae tendineae fused together and shortened Left atrium dilated.

38 Rheumatic Endocarditis of the Atrium —Involvement of the endocardium of the left atrium is frequent and is a well recognized lesion of rheumatic fever Irregular ridges and folds are found most commonly above the posterior leaflet of the mitral valve and may be continued across the atrial surface of this leaflet to the line of closure In other parts of the endocardium rounded or oval elevations may be seen These ridges and plaques are tawny yellow and dull in the acute stages as healing progresses they become gray and glistening and in some instances calcium is deposited in them Ulceration may occur over the calcium deposits These involved areas vary considerably in size they often measure several centimeters in greatest diameter or almost the entire endocardium of the atrium may be involved

tissue and covered by endothelium the vegetation is then healed and is gray translucent and smooth

The healing of these verrucae does not lead to the distortion of the valve leaflets *other changes that take place in the substance of the valves* are more important than the verrucae in causing distortion There is a diffuse inflammation characterized by polymorphonuclear leucocytes eosinophiles and large and small mononuclear cells (Fig 6) Aschoff nodules are occasionally found within the valve tissue The subsiding of this acute interstitial valvulitis is followed by an increase of connective tissue within the leaflet and the ingrowth of blood vessels at the base of the valve Palisades of large cells are at times to be found arranged at right angles to the surface and just below the endothelium covering the leaflet When the endothelium is damaged the vegetation forms

This acute interstitial valvulitis is doubtless repeated with each attack and following each attack the connective tissue is increased and vessels become more numerous within the valve The scar tissue becomes very compact the leaflet is thickened and calcium is often deposited in it Thick walled blood vessels at the base of the mitral leaflet and small collections of plasma cells and lymphocytes are found when the more acute reaction has subsided these cell accumulations persist for a long time The thickening fusion retraction and rigidity of the leaflets are the results of the interstitial valvulitis

The chordae tendineae are involved in a similar inflammatory process and tiny vegetations are sometimes found on them The healing of the interstitial inflammation leads to shortening fusion and beading of these structures The shortening of the chordae is frequently so marked that the tip of the papillary muscle is quite close to the margin of the leaflet and the chordae are fused to the ventricular surface of the valve leaflet

The fusion and retraction of the cusps lead to stenosis of the orifice and insufficiency of the valve (Fig 7) The orifice especially of the mitral valve may be considerably altered in shape and size Various terms such as buttonhole fish mouth and funnel are used to denote the deformities Large amounts of calcium may be found in the mitral and aortic leaflets As the leaflets of the aortic valve are shortened the edges seem to roll toward the sinuses of Valsalva consequently, if this rolling of a leaflet progresses far enough the central portion of a chain of verrucae may be seen on the sinus side of the leaflet The fusion of the aortic leaflets may be so extreme that it simulates a bicuspid valve



Fig 9 Rheumatic endocarditis of left atrium The endocardium is greatly thickened by fibrous tissue

are laid down in the scar and follow the course of the connective tissue The direction of these new elastic fibrils is opposite to that of the normal elastic fibrils of the atrial endocardium Not infrequently in the later stages of the lesion calcium is deposited

A somewhat different reaction is found in the outer less compact portion of the endocardium edema fibrin polymorphonuclear leucocytes and eosinophiles are present Aschoff nodules occur in this layer Granulation tissue from the adjacent myocardium penetrates into the outer part of the endocardium as the lesion heals

39 Rheumatic Pericarditis —The parietal and visceral pericardial surfaces may be entirely covered by fibrin or only a small area at the base may be involved The fibrin often has a ridged or irregular surface The amount of fluid in the pericardial space is variable The mesothelial cells may be preserved in small areas beneath the fibrin where they are swollen and some have multiple nuclei The exudate is replaced by granulation tissue that springs from the myocardium and parietal pericardium and may unite the two pericardial surfaces obliterating the pericardial space When the exudate is limited to the

The lesions vary *histologically* Swollen bands of collagen are surrounded in palisade fashion by large cells with pale vesicular nuclei (Fig 8) There are occasional Aschoff nodules though the elastic



Fig 8 Rheumatic endocarditis of left atrium There is marked infiltration by mononuclear cells into the endocardium These are collected in rows perpendicular to swollen collagenous fibrillae

fibers usually hold these cells in rows nodules are found also in the endocardium of the auricular appendage Collections of polymorphonuclear leucocytes with eosinophiles and large pale cells are present in other areas Mingled in these cell accumulations are cells with distorted or elongated nuclei some of these nuclei belong to polymorphonuclear leucocytes others to large mononuclear cells The elastic fibers are spread apart stretched fragmented or ruptured by these collections of cells

If the endothelium is damaged a vegetation forms on the surface This lesion heals with the ingrowth of an avascular connective tissue that is directed perpendicularly to the surface and connective tissue replaces any vegetation present (Fig 9) Later delicate elastic fibers

posited The elastic fibers are ruptured When the lesion heals scars without blood vessels extend into the subjacent media

In other areas involving the intima and at time the adjacent media collections of polymorphonuclear leucocytes large mononuclear cells and cells with distorted nuclei exactly reproduce the atrial lesion The long axis of the large cellular components is directed perpendicularly to the surface

Large basophilic cells are present in the media these are held in

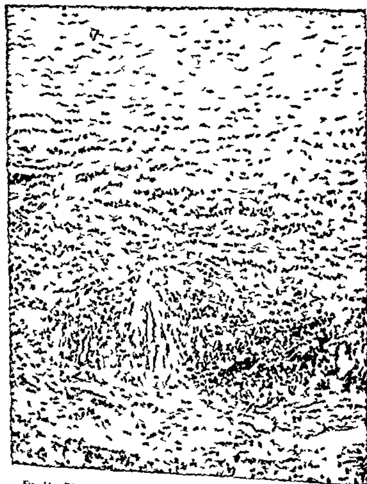


Fig 13 Rheumatic aortitis The inflammatory reaction involves the outer third of the media with a pronounced cellular infiltration by polymorphonuclear leucocytes Large mononuclear cells are held in rows by the elastic lamellae

base of the heart healing results in the formation of adhesions polypoid masses of connective tissue covered by mesothelium or in the proliferation of sheets of mesothelial cells

Aschoff nodules are occasionally found in the parietal pericardium

40 Rheumatic Aortitis and Pulmonary Arteritis —*Lesions in the aorta* are found that closely resemble those in the endocardium of the left atrium Similar lesions are found in the pulmonary artery Brownish yellow almost transparent elevated areas are to be seen in the intima these closely resemble in appearance the endocardial plaques and are quite easily distinguishable from those of arteriosclerosis or syphilis

*Histologically* the fibrillar material of the intima is swollen and about it collect large basophilic mononuclear cells arranged in a palisade manner (Fig 10) On the intimal surface some fibrin may be de

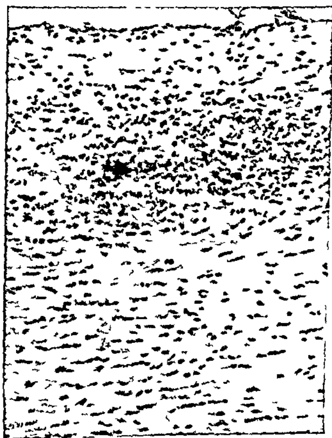


Fig 10 Rheumatic aortitis The lesion is limited to the inner third of the media and is characterized by a dense leucocytic infiltration

If only fibrin has been poured out beneath the endothelium it is replaced by connective tissue and the completely healed lesion then

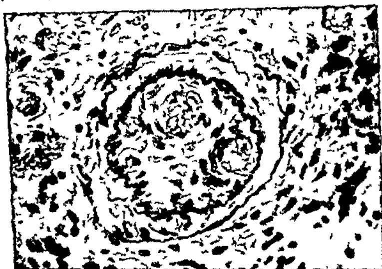


Fig 13 Rheumatic arteritis healed Th fibrin has been replaced by connective tissue and new channels have been formed

Fig 14 Verrucous endarteritis A mass of granular material covered by endothelium attached to the intima and almost fills the lumen

rows by the elastic fibers About the vasa vasorum are large cells and often with them are polymorphonuclear leucocytes and a few eosinophiles The nearby elastic fibers are fragmented though the fragmentation is not so great as that found in syphilitic aortitis (Fig 11) When the lesion heals there remains a dense avascular scar about the nutrient vessel Aschoff bodies are found in the adventitia or there may be single Aschoff cells at the junction of the media and adventitia With the healing of this lesion the adventitia is thickened by compact scar tissue Aneurysms due to rheumatic aortitis are rare

II RHEUMATIC ARTERITIS OF THE SMALLER ARTERIES —The involvement of the adventitia of the coronary arteries by Aschoff nodules is of frequent occurrence However these arteries are less often the seat of an acute inflammatory reaction due to rheumatic fever than are other small arteries as for example pulmonary renal and pancreatic arteries The acute lesion consists of the accumulation of fibrin beneath the endothelium and with the fibrin there may be hemorrhage The elastica interna is stretched fragmented or ruptured and the wall of the vessel is necrotic (Fig 12) Polymonphonuclear leucocytes

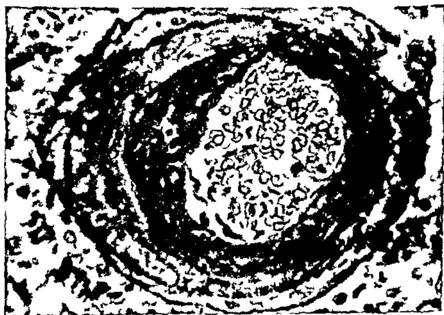


Fig 12 Rheumatic arteritis The acute lesion shows a thick layer beneath the endothelium the elastica is beaded and fragmented The lumen is patent

both neutrophilic and eosinophilic and large cells with distorted nuclei similar to those seen in the atrial endocardium collect about the vessel Thrombi in the lumen are extremely rare

If only fibrin has been poured out beneath the endothelium it is replaced by connective tissue and the completely healed lesion then

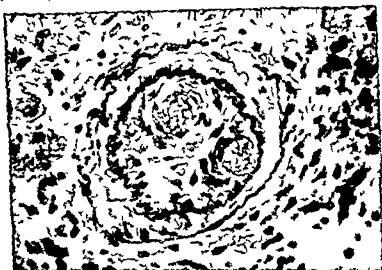


Fig 13 Rheumatic arteritis healed The *fibrin* has been replaced by connective tissue and new channels have been formed

Fig 14 Verrucous endarteritis A *mass* of granular material covered by endothelium is attached to the *intima* and almost fills the lumen

resembles an obliterating endarteritis However when there is hemorrhage and fibrin beneath the endothelium the endothelium extends down to surround the blood and form new capillaries that empty into the original but now narrowed lumen the fibrin is replaced by connective tissue The healed lesion when there have been hemorrhage and fibrin then has the appearance of an organized and canalized thrombus though no remnant of a thrombus is to be found nor is there any hemosiderin (Fig 13) This arteritis does not lead to aneurysm formation

Verrucous endarteritis is another specific type of arteritis that may affect the coronary arteries in rheumatic fever The lumen of the vessel contains a granular mass similar to that composing the verruca on the valve This granular mass is ultimately replaced by connective tissue and the surface is covered by endothelium (Fig 14)

## CHAPTER III

# SYPHILIS OF THE HEART AND AORTA

SYPHILIS of the cardiovascular system affects chiefly the aorta itself but the process may extend to the aortic valves to the tissues around the coronary orifices and rarely to the myocardium The pulmonary arteries are seldom involved

42 SYPHILITIC AORTITIS is most frequent in the ascending portion and arch less common in the thoracic part and in the abdominal segment The vessel wall is thickened and inelastic. The lumen is dilated

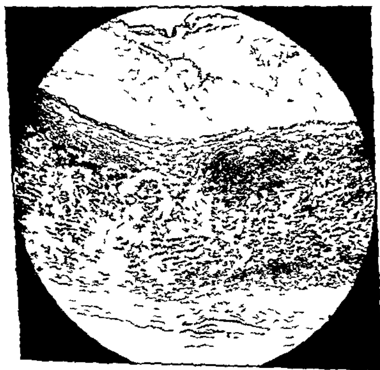


FIG. 15 Syphilitic aortitis Van Gieson and Weigert s elastic tissue stain The fibrous overgrowth of the intima is marked The media is considerably reduced in thickness and the continuity of dark staining elastica is interrupted in many places by irregular scars

and there may be fusiform or saccular aneurysms the latter occurring most often in the proximal portion The intima contains well defined pearly white hyaline elevations with smooth surfaces and of variable size Longitudinal wrinkling and dimpling of the intima are frequent The advancing edge of the lesion may be sharply demarcated Atheromatous deposits in the intima may complicate and obscure the appearance The intimal plaques may encroach upon and obstruct the orifice of any arterial branch including those of the coronary arteries

*Histologically* (Figs 15 and 16) there is increase of connective tissue in the adventitia thickening and endothelial proliferation in the vasa vasorum and perivascular infiltration of lymphocytes and plasma cells The media is interrupted by scars and penetrated at intervals by



Fig 16 Syphilitic aortitis There is a chronic inflammation of the media and of the thickened adventitia which shows in the lower part of the photograph Many small foci of lymphocytes and plasma cells are scattered throughout In one area indicated by the arrow a portion of the media has undergone gummatous necrosis

thin walled vascular channels and collections of lymphocytes and plasma cells usually situated in areas of scarring where the elastic lamellae are disrupted and the smooth muscle cells are replaced by collagen fibers Occasionally the larger cell aggregates show miliary gummata with central areas of necrosis Rarely a few multinuclear foreign body giant cells may also appear The intima is irregularly thickened by new connective tissue Treponema pallida cannot usually be demonstrated in the lesions although their presence has been described

In some instances of clinical cardiovascular syphilis with gross features of aortitis the inflammatory features may be entirely absent histologically

43 Syphilitic Aneurysm of the Aorta—Syphilitic aneurysms of the thoracic aorta may be saccular or fusiform single or multiple. The commonest sites in order of frequency are (1) ascending (2) transverse and (3) descending thoracic aorta Aneurysms of the abdominal aorta of syphilitic origin are rare

Saccular aneurysm is the commoner type and may vary greatly in size It is usually connected with the lumen of the vessel by a single well formed stoma Its lining may be smooth wrinkled ulcerated or covered by a thrombus The latter may be laminated by successive deposits of blood elements At the junction of thrombus and sac wall evidence of organization may be seen There may be several aneurysms of variable size and age in a single aorta The adjacent structures are subject to erosion compression obstruction or displacement More than one half of all such aneurysms rupture Rupture leads to hemorrhage the distribution of which depends upon the site of the tear Spontaneous rupture of the aorta in syphilitic aortitis without aneurysm has been described but is extremely rare

Fusiform or diffuse aneurysmal dilatation of the aorta due to syphilis is also common in the thoracic portion and may be associated with saccular aneurysm in some other part Its extent is variable Thrombosis in this type is rare

*Histologically* the wall of a sac is composed of dense hyalinized connective tissue which sometimes becomes calcified and even ossified The layers of the original vessel wall may be lacking and the specific character of the syphilitic lesion obscured except at its mouth or in sections from the adjacent aortic wall The sac is lined by endothelium or organizing thrombus In fusiform aneurysm remnants of the original vessel wall usually persist and syphilitic changes may still be detectable

44 Syphilitic Involvement of the Aortic Valve is found in one third of all cases of syphilitic aortitis The valve leaflets are cicatrized and incompetent as a result of inflammatory change extending in from the underlying aortic wall The lateral parts of the aortic cusps may coalesce with the intima of the aorta leading to separation of the *commissures and incompetency with exposure of the sinuses of Valsalva* Hyaline plaques in these areas denote underlying commissural scars The margins of the cusps in more advanced cases show thickening and rolling toward the sinus aspect Further cicatrization may reduce the cusps to cordlike structures and increase their incompetency Dilatation of the aortic ring and supravalvular portion of the aorta without changes in the cusps also may lead to incompetency of the valve

Isolated syphilitic valvulitis is extremely rare but gummata are occasionally seen at the point of attachment to the aortic wall

Rarely the anterior leaflet of the mitral valve is affected by extension from syphilitic aortitis but the leaflet is not uncommonly thickened in the presence of aortic insufficiency without showing evidence of syphilis

*Microscopic examination* of the affected leaflets and of the aortic ring and root of the aorta shows endothelial proliferation in the vasa vasorum and perivascular infiltration by lymphocytes and plasma cells in the adventitia of the aorta near the ring in addition to productive changes

45 Coronary Artery Involvement in Syphilitic Aortitis frequently causes stenosis or almost complete obliteration of the orifices particularly those which arise abnormally high Complete occlusion rarely occurs Syphilitic coronary arteritis unassociated with syphilitic aortitis is rare

*Microscopic sections* of the stenosed portion of such coronary arteries usually reveal intimal fibrous proliferation The site of the original lumen may be demonstrated only by elastic tissue stains In rare cases there is continuity of the syphilitic process in the aorta and that of the proximal part of the artery The thickened intima and even the media may contain granulation tissue Overgrowth of the adventitial connective tissue is also present as well as obliteration of the vasa vasorum and perivascular infiltration by lymphocytes and plasma cells

46 The Myocardium in Syphilis—The main effects of syphilis on the myocardium are produced by lesions in the aorta particularly coronary ostial stenosis Gummata solitary or multiple have been



Fig. 17. Low power view of a section of gumma at its myocardial edge shows the muscle bundles terminating at the edge of a necrotic acellular zone.



Fig. 18. Gumma of myocardium. Granulation tissue present at margin of gumma; also multinucleated giant cells, lymphocytes, plasma cells and large mononuclear cells.

described but are rare They may be found anywhere but are most frequent in the wall of the left ventricle near the base or in the interventricular septum In the latter site they sometimes involve the bundle of His and cause complete heart block Rarely they may result in aneurysm formation Diffuse interstitial myocarditis has been noted in patients dying in the secondary stage of syphilis These instances are so rare as to be considered curiosities

*Microscopically* myocardial gummata (Figs 17 and 18) replace the muscle bundles or produce pressure atrophy of adjacent muscle fibers

47 CONGENITAL SYPHILIS may involve the heart and great vessels in infants who are stillborn die shortly after birth or live for an indefinite period Spirochetes are frequently present in large numbers with or without anatomical lesions They are unusually abundant in the macerated fetus Myocarditis coronary arteritis and aneurysm have been recorded In myocarditis there may be edematous areas overrun with spirochetes Associated with these lesions there may be fibrosis of the endocardium

CHAPTER IV

# DISEASES OF THE CORONARY ARTERIES

48 ARTERITIS OF THE CORONARY ARTERIES (endarteritis mesarteritis periarteritis) may be found in such diseases as typhus fever diphtheria scarlet fever rheumatic fever pneumonia and septicemia or may be part of a generalized vascular reaction Inflammation may involve the intima primarily with secondary extension to the media More rarely all coats may be involved resulting in abscess or mycotic aneurysm sometimes with rupture and hemorrhage Periarteritis may be seen when abscesses of the myocardium or suppurative pericardial lesions are in proximity to the coronary vessels In these cases the lesion may eventually involve all coats of the arterial wall

The *microscopic picture* varies depending upon the causative agent i e toxins bacteria or emboli The injury to the vessel wall may be slight or severe

The lesion attributed to the toxins of infectious agents is usually evidenced by endothelial proliferation and by the presence of fat in the injured cells less often by fibrinoid change or by *necrosis* In lesions due to bacteria the response is usually suppurative Necrosis with lack of cellular response is also seen in some lesions and *mycotic* aneurysms may ensue Frequently an embolus containing bacteria may occupy the lumen Occasionally a secondary thrombus may surround the original embolus while in other instances thrombosis may be incidental to the arteritis Micro-organisms should be identified when possible

49 PANARTERITIS NODOSA (polyarteritis nodosa periarteritis nodosa) is an inflammatory disease in which no specific etiological agent has been demonstrated but is considered to be a hyperergic inflammation of the arterial wall *The disease affects the medium-sized arteries* The veins are not involved The secondary branches of the main coronary arteries are often affected

The outstanding features of the disease are inflammation of all coats with conspicuous medial and adventitial involvement and perivascular infiltration leading occasionally to the formation of visible nodules Thrombosis of the coronary arteries with infarction of the myocardium

and aneurysm or rupture of a necrotic artery with hemorrhage into the adventitia or epicardium may occur These lesions are readily seen on gross inspection

*Histologically* (Fig 19) in the acute stage there is dense infiltration

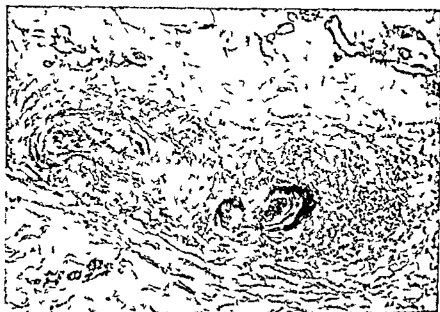


Fig 19 Acute periarteritis nodosa The adventitial cellular infiltration is pronounced The media of one vessel is necrotic and dark staining The lumina are still patent

of the media and adventitia by polymorphonuclear neutrophiles eosinophiles lymphocytes and plasma cells The elastic lamellae are frequently fragmented or destroyed Fibrinoid degeneration and necrosis in the medial and adventitial layers are characteristic features The intima is frequently involved and secondary thrombosis of the lumen may occur The periarterial connective tissue may become edematous and infiltrated by leucocytes and erythrocytes Later there is proliferation of fibroblasts from the adventitia into the inflammatory zone with an increase in lymphocytes plasma cells and sometimes eosinophiles in large numbers Granulation tissue and subsequently connective tissue replace the destroyed components of the wall with ultimate canalization of thrombi

Aneurysm a rather common sequel of segmental destruction of the arterial wall in association with periarterial cellular infiltration or connective tissue proliferation may produce visible nodules In the majority of cases both fresh and healed lesions occur together However occasionally only a healed stage of periarteritis nodosa is found

Scars in the myocardium representing healed infarcts may be the only naked-eye finding The vessels may be tortuous and studded with bluish white nodules not unlike those seen in acute stages of the disease The lumina may be greatly reduced or obliterated Periarterial mantles of scar tissue are characteristic of the end stage

*Histologically* the healed stage (Fig 20) is characterized by connec



Fig 20 Healed panarteritis nodosa with aneurysm

tive tissue replacement of the arterial wall with marked stenosis of the lumen The intima may be variously deformed depending upon previous thrombosis organization and canalization or there may be simple fibrous thickening There is interruption of elastic fibers by scars in the media The vessel appears embedded in a dense mass of contracted and occasionally hyalinized connective tissue Occasionally sections may include small aneurysmal pouches

50 Thromboangiitis Obliterans of the Coronary Arteries —Although this disease of unknown cause usually affects the vessels of the extremities a few authentic reports of coronary involvement are on record Advanced coronary arteriosclerosis with thrombosis occurring in patients with thromboangiitis obliterans elsewhere may present

difficulties in differential diagnosis To justify the diagnosis features of the lesion in the coronary vessels should correspond with those in the vessels of the extremities The myocardium may show recent or old infarcts

*51* ARTERIOSCLEROSIS is the commonest affection of the coronary arteries and is characterized by variable degrees of intimal changes These consist of lipid deposition collections of cholesterol bearing macrophages eccentric or concentric fibrotic thickening foci of calcification and occasionally bone formation Frequent accompaniment of arteriosclerosis is elastic fragmentation and medial atrophy which appears to be *proportional to the thickness of the adjacent* intimal plaque Periarterial lymphocytic infiltration is common Associated with the arteriosclerotic intimal thickening there is often a rich capillary formation which originates usually from the vasa vasorum and to a lesser degree from the endothelium of the coronary artery *proper* (Fig 21) These intimal capillaries are the sequel of the degenerative

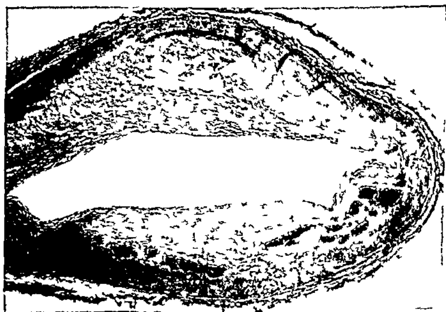


Fig 21 Photomicrograph showing arteriosclerotic plaque with many capillaries in thickened intima (Courtesy of H Horn and L E Finkelstein)

and proliferative changes incident to arteriosclerosis The picture of arteriosclerosis consists primarily of intimal lipoidosis and collagenous thickening resulting in intimal fibrosis *brought about with the aid of* new capillary channels The latter proliferate in association with fatty

change and intimal thickening Necrosis of the plaque may result in an atheromatous abscess (Fig 21) Commonly associated with degenerating foci within arteriosclerotic plaques are areas of recent hemorrhage and deposits of iron pigment incident to rupture of intramural capillaries and disintegration of hemoglobin (Fig 22) Such hemorrhage

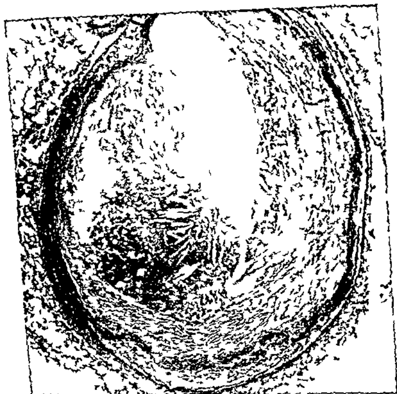


Fig Recent hemorrhage into atheromatous plaque (Courtesy of H Horn and L. E Finkelstein)

frequently acts as the precipitating factor in coronary artery occlusion

Acute coronary artery occlusion may be effected (1) by focal disruption of the intima (2) by hemorrhage into an atheromatous abscess causing compression of the lumen (Fig 22) (3) by thrombosis through the mechanism of recent intimal and endothelial changes induced by a remote focus of intimal hemorrhage (4) by endothelial injury resulting from the impingement of an atheromatous abscess and associated hemorrhage (Fig 23) and finally (5) thrombi may be

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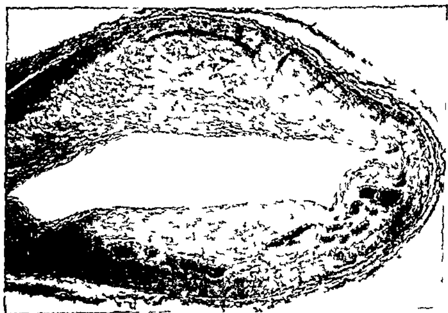


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implanted on arteriosclerotic lesions which have narrowed the lumen of the vessel (Fig 24) It is important to observe the morphogenesis of coronary thrombi their age size and extent as well as the condition of the arterial wall When two or more thrombi are present histological s ctions may reveal different pictures varying from recent or partially organized thrombi to healed recanalized lesions The latter are frequently difficult to distinguish from occlusions which have resulted from slowly progressive arteriosclerosis

The most severe changes usually are found in the proximal portions of the left coronary artery and its anterior descending branch the right coronary artery and the circumflex branch of the left somewhat less frequently present advanced changes Extensive arteriosclerosis with calcification and associated stenosis of the lumen is commonly found in several major branches of the coronary artery Almost total obliteration of the lumen is frequent Because of the likelihood of multiple areas of stenosis all coronary arteries should be examined even after one advanced lesion has been demonstrated Pronounced changes may occur in the more distal parts especially at bifurcations The coronary arteries should be examined in their entirety by frequent transverse sections at intervals of 2 millimeters to 3 millimeters The severity of the lesion at various levels its effect on the size of the lumen its nature and location should be described

Routine microscopic examination should be done in all instances of coronary arteriosclerosis even in the absence of naked-eye myocardial alteration This is essential to detect lesions of microscopic extent Variation in the character and intensity of the arteriosclerotic process in the same artery may range from simple yellow discoloration to fibrotic or calcified plaques with foci of degeneration or to an organized and canalized thrombus occluding the vessel lumen (Fig 25) Many histologic sections are necessary

Coronary thrombosis occurs almost always in association with arteriosclerosis It may occur occasionally in coronary arteritis and more rarely is secondary to an embolus

A coronary thrombus may be recent (acute fresh subacute) or old (organized healed) The former is usually red or grayish red the latter gray or grayish white Fresh thrombi may be dislodged from the vessel wall unless care is exercised Organized thrombi are firmly adherent and differentiation from the arterial wall may be difficult

Although thrombi may be located anywhere the commonest sites are (1) in the anterior descending branch of the left coronary artery about 2 centimeters to 3 centimeters distal to its orifice (2) in the right circumflex branch within 3 centimeters to 5 centimeters of its origin

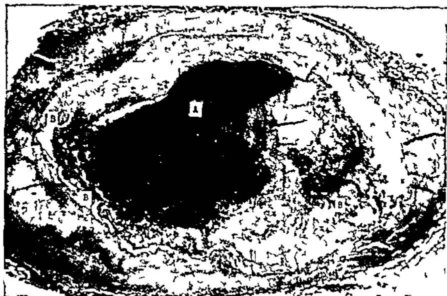



Fig 23 Coronary arteriosclerosis Hemorrhages (B) in atheromatous plaque with thrombus (A) in lumen (Courtesy of H Horn and L E Finkelstein)

Fig 24 Organizing thrombus of a coronary artery

52 ANEURYSMS OF THE CORONARY ARTERIES are rare There are two main groups (1) mycotic-embolic and (2) arteriosclerotic. Syphilitic arteritis is an occasional underlying cause The mycotic-embolic type may be secondary to bacterial endocarditis Aneurysms of the coronary arteries are usually single They vary in size from one to several centimeters in diameter Rupture has taken place in approximately one half of those observed at autopsy The proximal part of the left coronary artery is most often involved

*Microscopically* fragmentation of the elastic and muscle fibers of the media occurs Foci of necrosis may be seen in the intima and media or the entire wall may be necrotic Aneurysms formed on a mycotic embolic basis may show partial or complete occlusion of the lumen by a plug in various stages of organization or by acute inflammation of the vessel wall In the majority of arteriosclerotic aneurysms an advanced degree of arteriosclerosis is found occasionally with dissection of the wall by hemorrhage

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54 RUPTURE (spontaneous) OF THE CORONARY ARTERIES is rare Mycotic-embolic aneurysms and especially the aneurysms that accompany panarteritis nodosa may induce rupture of the arterial wall Rarely rupture of the wall due to an arteriosclerotic dissecting aneurysm may be seen The histological findings are those of the underlying disease i e arteriosclerosis arteritis panarteritis nodosa



FIG 25 Organized thrombus of a coronary artery It is difficult to determine how much of this occluding mass represents intimal plaque formation and how much organized thrombus

(3) in the left circumflex branch just beyond its origin from the main trunk

The majority of coronary artery occlusions by thrombosis are associated with hemorrhages into the intima The thrombi are usually short in extent longer ones may form by propagation *Multiple thrombi* in the coronary arteries or even in one branch are not unusual They may be of different ages varying from fresh to organized ones This again emphasizes the need for diligent gross examination

The method of choice for the examination and study of the coronary arteries is by transverse section A more intricate method is the injection of the coronary circulation with a gelatin suspension of barium with or without a pigment followed by roentgen examination and transverse sectioning *

*M J Schlesinger *American Heart Journal* 15 528 1938

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CHAPTER V

# DISEASES OF THE MYOCARDIUM

55 ACUTE MYOCARDITIS—Focal or diffuse interstitial myocardial inflammation may occur in some infectious diseases apart from endocardial or other changes in the heart produced by the localization of bacteria It may occur in prolonged septic states after exanthemata in some virus and rickettsial infections (Fig 26) occasionally in uremia



Fig [illegible] An example of rickets infecti on showing fibrinoid degenerati n in an affected small coronary arterial wall and in the peri arterial supporting [illegible] th focal myocard al degeneration and some interstital infiltration by le icocytes and histi cytes This lesion is encountere l in epidemic typhus fever and spotted fever (Courtesy of Armed Forces Institute of Pathology)

in other debilitating conditions and after drug or foreign serum therapy Lesions may be found in the muscle arterioles and in the perivascular supporting tissues

Grossly the heart may be flabby or normal pallid or yellowish with dilated chambers In severe instances mural thrombi may be present.

*Microscopically* there are interstitial infiltrations of histiocytes lymphocytes and eosinophilic or neutrophilic polymorphonuclear leu

cocytes between the muscle fibers and in the perivascular connective tissue In severe instances the muscle may exhibit fatty granular or hyaline degeneration In subacute bacterial endocarditis apart from focal embolic lesions focal intrafascicular infiltrations of inflammatory cells may occur

56 ABSCESSES OF THE MYOCARDIUM rarely macroscopic are encountered in bacteremia Fresh lesions are usually found in relationship to terminal branches of the coronary arteries the necrotic walls of which may be still visible the whole surrounded by polymorphonuclear leucocytes Bacterial stains may be necessary to differentiate abscesses from focal necroses of the ischemic type The Staphylococcus is the commonest organism found in these cases

57 IDIOPATHIC MYOCARDITIS (Fiedler's myocarditis isolated myocarditis interstitial myocarditis acute or subacute productive myocarditis fibrous myocarditis) is a form of cardiopathy of unknown cause occurring chiefly in adults principally in males who present cardiac enlargement and diffuse inflammatory changes limited to the myocardium The coronary arteries may be normal In the early stages the heart may present softening areas of fatty change and zones of hyperemia Intraventricular thrombi may be attached to adjacent areas of myomalacia later scars may be visible throughout the myocardium

*Microscopic* examination may reveal disseminated necrosis of the myocardium together with marked fixed tissue response and areas of replacement fibrosis Wide bands consisting of lymphocytes histiocytes plasma cells and sometimes numbers of eosinophiles may accompany these lesions Muscle giant cells may be prominent

58 ATROPHY OF THE HEART results from diminution in the size of its muscle fibers

59 BROWN ATROPHY is characterized by atrophy and pigmentation of the muscle fibers with diminution in the size of the heart It is found in starvation cachexia and senility Occasionally it is associated with serous atrophy of the epicardial fat The lipochrome pigment is deposited in the sarcoplasm at both ends of the nuclei

60 FAT INFILTRATION OF THE MYOCARDIUM—Excessive deposits of adipose tissue may occur among muscle bundles especially in obese individuals In them the epicardial fat may be increased encasing the entire heart

On section the fat deposits may extend between and replace muscle bundles most often in the right ventricle The fat may be visible through the endocardium Widely separated muscle fibers or bundles sometimes undergo pressure atrophy

61 Fatty Degeneration of the Myocardium may occur in all severe anemias The heart may be enlarged dilated flabby and often presents transverse yellow markings due to fatty change visible through the endocardium These are variously designated as tabby cat or tiger markings Special fat stains reveal rows of fat droplets in the muscle fibers

62 Toxic Myocardial Degeneration —The myocardium is susceptible to systemic diffusible toxins or chemicals which produce softening Among the latter carbon tetrachloride chloroform phosphorus arsenic and carbon monoxide are most often encountered These degenerations result in reparative processes of considerable degree and may be seen in other toxic states particularly in carbon monoxide poisoning Myocarditis associated with diphtheria is an example In the early stages minute focal extravasations of blood may occur Otherwise the lesion is only found histologically (Fig 27)



Fig Myocardium in diphtheria Highly magnified photomicrograph showing necrotic myofibrils and histiocytic and leucocytic infiltration as well as edema

63 MYOCARDIAL DEGENERATION IN FRIEDREICH'S ATAXIA —Myocardial involvement without pericardial or valvular disease has been found frequently in *Friedreich's ataxia*. *It consists of cardiac hypertrophy* with focal destruction of muscle fibers with fibrosis and hypertrophy of intact fibers. Focal interstitial myocarditis characterized by lymphocytic infiltration and occasional polymorphonuclear leucocytes may also be observed.

64 CALCIFICATION OF THE MYOCARDIUM —Primary calcification is rare. Calcium compounds may be deposited in freshly infarcted and necrotic muscle fibers in scars or in other forms of myocardial change. *Metastatic calcification is seen in hyperparathyroidism or other forms* of severe osteoporosis. It may be found in chronic renal disease and in hypervitaminosis D.

65 GLYCOGENOSIS (von Gierke's disease, glycogen infiltration) —This is a rare lesion which may be encountered in infants. It is characterized by cardiac enlargement and hepatomegaly. Excessive glycogen deposits are demonstrable in the affected organs without any associated inflammatory process. Some of the cases of so called "congenital idiopathic cardiac hypertrophy" may represent instances of this disturbance.

66 AMYLOID INFILTRATION OF THE HEART is usually associated with more extensive deposits in other organs. In persons over 80 years of age it may be found as a primary and isolated lesion. It is often deposited in the interventricular septum. The myocardium becomes pale, smooth, firm and waxy. In most instances the heart remains normal in size. The amyloid material is found in the capillary walls and interstitial tissues. The muscle fibers undergo gradual pressure atrophy and in some cases large portions of the myocardium may be replaced.

67 FIBROSIS OF MYOCARDIUM (replacement fibrosis, myocardial fibrosis) —Under this term are included changes due to diminished nutrition *of the muscle fibers without demonstrable infarction* and with resultant fibrous replacement. The cause is usually coronary arteriosclerosis with gradual stenosis. More rarely syphilitic ostial stenosis is the cause.

Fibrosis of the myocardium is usually patchy and may be seen as white or gray strands or focal scars in the *left* ventricle, occasionally in the right one, and rarely in the atria. Hearts which most often show myocardial fibrosis are those with severe coronary arteriosclerosis. Less

frequently hearts with marked hypertrophy associated with arterial hypertension and only slight or moderate coronary arteriosclerosis may exhibit myocardial fibrosis

*Microscopically* (Fig 28) the important differential feature is its involvement of the muscle as opposed to the perivascular distribution



Fig 28 Replacement fibrosis of the myocardium There is irregular scarring of the myocardial bundles which is both perivascular and interstitial in distribution

of the scars of rheumatic fever Residual inflammatory cell collections are not seen

Examination of the coronary arteries frequently reveals moderate to advanced degrees of arteriosclerosis endarteritis or thrombotic occlusion

68 INFARCT OF THE MYOCARDIUM —An infarct of the heart may be either a focal and microscopic or a massive lesion leading to thinning of the ventricular wall aneurysmal bulging or even to rupture

The size and number of infarcts are dependent upon the degree and

distribution of coronary arteriosclerosis and upon other factors which may induce coronary insufficiency

Pulmonary embolism severe acute anemia prolonged traumatic shock and carbon monoxide poisoning may result in myocardial ischemia by reducing coronary flow or the nutrition of the myocardium to the point where infarction may ensue Hypertrophied hearts are particularly vulnerable to reductions in effective coronary flow Certain valvular defects especially aortic stenosis and insufficiency, when associated with severe prolonged disturbances in cardiac rate and rhythm may *lead to myocardial ischemia and focal necrosis in the absence of* coronary obstruction Such lesions may be focal and disseminated Most infarcts that attract notice are massive and follow coronary arterial occlusion Areas of the myocardium least supplied with collateral channels are particularly vulnerable to coronary insufficiency

Most infarcts of the heart are found in the left ventricle in its anterior and apical portion somewhat less often in the basal portion of the posterior wall or in the median portion Not infrequently two or more of these sites are involved There is a high incidence of occlusion either by a thrombus or by extreme stenosis in the anterior descending branch of the left coronary the right coronary or the circumflex branch of the left coronary The underlying disease is usually arteriosclerosis infrequently syphilitic aortitis at the root of the aorta A rare cause of infarction is occlusion by embolism

In infarcts of the anterior wall of the left ventricle the anterior part of the interventricular septum and the apex are frequently included This area corresponds to the distribution of the anterior descending *branch of the left coronary artery* The *posterior portion* of the septum is often involved in infarction of the corresponding wall of the left ventricle caused usually by obstruction of the right coronary artery Infarction of the more median part of the left ventricular wall results from closure of the circumflex branch of the left coronary artery The right ventricle and the atria are rarely the seat of infarction Multiple infarcts recent or healed or combinations of both in the same area are not uncommon

For diagnosis multiple sections not more than 1 centimeter in thickness should be made transversely or sagittally *through the myocardium* of the ventricles Another requisite is detailed examination of the coronary arteries (Chapter IV)

Infarcts may be recent (acute fresh hemorrhagic necrotic subacute) or *old* (*healed* ancient organized) An infarct may be recognized by its color and consistency Dark red with yellowish mottling is the usual coloring of fresh infarcts The tissue is softer than normal so that not

infrequently the area involved appears sunken when seen from the epicardial surface Its appearance varies with its age Weeks after the onset it presents a mottled grayish red or yellowish appearance the former due to connective tissue proliferation the latter to fatty changes

Healed infarcts are readily seen if they extend to the endocardium They appear as slightly depressed shiny white or grayish white scars of varying size Healed infarcts in contrast to fresh infarcts often bulge above the epicardial surface with or without a localized patch of adherent pericardium Those occurring in the posterior wall of the left ventricle may be easily overlooked because they usually lie behind the posterior leaflet of the mitral valve or its papillary muscles Occasionally a healed infarct is situated in the wall of the ventricle between the two surfaces and is detected only by transverse or sagittal sections

The size of the infarct may vary from a barely visible size to one that is 10 centimeters or more in maximum width The lesion may or may not involve the pericardium or endocardium where it may set up either a localized or diffuse fibrinous reaction It may be followed by adhesive pericarditis or on the endocardium there may be a thrombus As a rule the intraventricular thrombus is seen in the left ventricle When infarction has extended completely through the interventricular septum a mural thrombus may also be found in the right ventricle

The histological findings depend upon the age of the infarct In the early stages interstitial edema congestion of small blood vessels pyknosis of nuclei some loss of cross striations and necrosis of muscle fibers are constant This is followed by complete necrosis of the muscle cells extravasation of red blood cells and polymorphonuclear leucocytic infiltration Phagocytosis of necrotic cells and nuclear elements is seen early and fatty metamorphosis becomes apparent in the necrotic and degenerated fibers The sarcoplasm becomes homogeneous dense deeply eosinophilic and loses cross striation In the immediate vicinity frequently there are pools of deeply basophilic material generally believed to be nucleoprotein derived from the dead cells Fibroblastic proliferation is usually conspicuous by the second week and there is diminution in the inflammatory response although considerable necrosis may still be seen By the third or fourth week areas of fibrosis are visible and are interspersed by richly vascularized granulation tissue By the third or fourth month repair and scarring usually are complete (Fig 29) Occasionally only the persistence of blood pigment in clusters of macrophages suggests a former infarct In late stages cal

cification may appear in the infarcted area or in the overlying thrombus

69 Aneurysmal Dilatation of the Heart (aneurysm of the heart, cardiac aneurysm) usually occurs in the wall of the left ventricle



Fig 29 Healed infarct of myocardium The necrotic muscle has been replaced by fibrous tissue which ramifies irregularly In places it encloses groups *of living muscle bundles The scar tissue is relatively acellular but still con*tains numerous thin walled blood channels Note the loss of bundle outlines

Aneurysms vary in size from a few centimeters to such as involve half of a ventricle A thrombus is often adherent to its wall and may partially or even completely fill the pouch In many instances however the lining is white smooth or corrugated and glistening *Pericardial* adhesions often form

*Microscopically*, the wall of the aneurysm usually consists of dense

scar tissue Occasionally bands of partially preserved muscle fibers may be seen The endocardium is often thickened

70 RUPTURE OF THE MYOCARDIUM SPONTANEOUS —From 2 to 9 per cent of all cases of myocardial infarcts both recent and old rupture A cardiac aneurysm is occasionally the site of rupture and rarely a gumma

Ruptures usually occur in the anterior or posterior wall of the left ventricle More rarely other chambers the interventricular septum or a papillary muscle may rupture As a rule the site of rupture is in the center of a recent infarct and usually occurs during the early stages after onset Less frequently rupture of a healed infarct with or without superimposed fresh infarction is seen The tear may vary in length and width from a few millimeters to several centimeters No characteristic appearance is shown by the line of rupture Some are ragged and irregular resembling traumatic ruptures some cleanly cut and regular like incised wounds and others pursue a tortuous course through the heart wall with small external and internal openings

*Histologically* the lesion in most cases presents a central area of hemorrhage and necrosis in the center of which a linear tear may be seen

71 ENLARGEMENT OF THE HEART is one of the commonest abnormalities Since the chief criterion is the weight it is essential to know the normal variation There is a definite correlation between heart weight and body weight The weight of the heart increases with body weight irrespective of age The average weight of the adult male heart is 300 grams and of the female 250 grams The accompanying revised table may be used as a guide In determining the weight care should be taken to remove clots from the chambers and to note whether or not the aorta is included

Enlargement of the heart may be due to hypertrophy of muscle or dilatation of chambers or more commonly to a combination of both A hypertrophied heart may dilate yet gross and microscopic examination may reveal no evidence of other myocardial disease

The two most common causes of hypertrophy are arterial hypertension and valvular deformities However there are instances when the heart weight is not increased in these conditions Other less frequent causes are renal hypertension chronic pulmonary disease (bronchiectasis emphysema with chronic interstitial pneumonitis and silicosis) myocardial infarcts congenital cardiac anomalies severe anemia

H L Smith *American Heart Journal* p ?? October 19?8

## NORMAL WEIGHT OF THE HEART

(Male and Female)

| Body Weight | | | | Minimum Gm | | Average Gm | | Maximum Gm | |
|---|---|---|---|---|---|---|---|---|---|
| Pounds | | Kilograms | | | | | | | |
| M | F | M | F | M | F | M | F | M | F |
| 105 | 90 | 47 | 40 | 165 | 135 | 205 | 162 | 244 | 193 |
| 110 | 95 | 50 | 43 | 173 | 143 | 215 | 171 | 253 | 204 |
| 115 | 100 | 52 | 45 | 181 | 150 | 225 | 180 | 264 | 215 |
| 120 | 105 | 54 | 47 | 190 | 158 | 235 | 189 | 276 | 226 |
| 125 | 110 | 56 | 50 | 198 | 165 | 245 | 198 | 287 | 237 |
| 130 | 115 | 58 | 52 | 206 | 172 | 255 | 207 | 299 | 248 |
| 135 | 120 | 60 | 54 | 213 | 180 | 265 | 215 | 310 | 259 |
| 140 | 125 | 63 | 56 | 221 | 188 | 274 | 225 | 322 | 268 |
| 145 | 130 | 65 | 58 | 229 | 195 | 284 | 234 | 333 | 277 |
| 150 | 135 | 68 | 60 | 237 | 203 | 294 | 244 | 345 | 286 |
| 155 | 140 | 70 | 63 | 245 | 211 | 304 | 253 | 356 | 295 |
| 160 | 145 | 72 | 65 | 253 | 219 | 313 | 262 | 368 | 304 |
| 165 | 150 | 74 | 68 | 261 | 225 | 323 | 272 | 370 | 313 |
| 170 | 155 | 77 | 70 | 268 | 233 | 333 | 282 | 371 | 322 |
| 175 | 160 | 79 | 72 | 280 | 240 | 343 | 288 | 372 | 330 |
| 180 | 165 | 81 | 74 | 288 | 247 | 353 | 297 | 374 | 337 |
| 185 | 170 | 83 | 77 | 296 | 255 | 363 | 306 | 382 | 343 |
| 190 | 175 | 86 | 79 | 304 | 283 | 373 | 315 | 392 | 350 |
| 195 | 180 | 88 | 81 | 312 | 301 | 382 | 324 | 402 | 356 |
| 200 | 185 | 90 | 83 | 320 | 309 | 392 | 333 | 412 | 361 |
| | 190 | | 86 | | 317 | | 342 | | 366 |
| | 195 | | 88 | | 325 | | 351 | | 371 |

and hyperthyroidism Among the rare etiological factors are deformities of the thorax especially kyphoscoliosis glycogen storage disease arteriovenous communications (congenital or acquired) myxedema beriberi hemochromatosis and idiopathic myocarditis

In about 10 to 20 percent of hypertrophied hearts the etiology may not be *determinable* A *certain proportion of these* however present coronary arteriosclerosis with patchy fibrosis or healed infarcts in the myocardium One group may present endocardial fibrosis in plaques with or without overlying mural thrombi others are the seat of various types of degeneration or inflammation

72 HYPERTROPHY OF THE MYOCARDIUM —The myocardium is usually firm and dark reddish brown Most hypertrophied hearts show some dilatation Hypertrophy is frequently limited to one chamber particularly the left ventricle or two may be involved as in mitral stenosis with left atrial and right ventricular hypertrophy The entire heart however often participates

The weight and relative sizes of the chambers should always be noted This may be done by comparing the sizes of the atria and ventricles The thickness of the ventricle walls at the base at the midpoint between apex and mitral ring on the left side and apex and pulmonary valve on the right side and finally at the apex should be recorded Erroneous figures are apt to be obtained if the papillary muscles and columnae carneae are included in the measurements

*Histologically* the principal finding is hypertrophy of muscle fibers without increase in number The change is especially noticeable in the increased average transverse diameter of the muscle cells There is also some increase in the length of the fibers due to increase in sarcoplasm as well as myofibrils The nuclei are enlarged with blunt ends and may be hyperchromatic. There is an inadequate capillary blood supply to the hypertrophied myocardial fibers because of failure of the capillaries to increase in number proportionately

If the hypertrophy is due to intrinsic disease evidences of myocardial damage either degenerative or inflammatory are coexistent Patchy fibrosis or healed infarcts may replace muscle tissue Other hearts may show perivascular scarring with or without residual cell collections or *more rarely diffuse* fibrous changes representing the end stage of an inflammatory process Idiopathic (isolated) myocarditis is an example of hypertrophy resulting from the response of the heart to diffuse inflammation

73 DILATATION OF THE MYOCARDIUM —When dilatation is predominant the heart presents a *globoid* form The muscle is flabby the chambers enlarged There is flattening of the papillary muscles and trabeculae carneae of the ventricles and of the pectinate muscles when the atria are involved

*Microscopically* there are various types of degeneration inflammation or focal necrosis depending upon the cause An important diagnostic feature of dilatation is the conspicuous appearance of the intercalated discs especially in the muscle fibers near the endocardium

74 GRANULOMATA OF THE MYOCARDIUM INFECTIOUS —a In syphilis the myocardium is rarely involved (Chapter III)

b Tuberculosis is rare It is most frequently encountered as microscopic miliary tubercles associated with generalized miliary tuberculosis or as an extension from tuberculous mediastinal lymph nodes In tuberculous pericarditis the inflammation may sometimes spread to the myocardium Rarely nodular tuberculomata form and many simulate gummata Tuberculosis of the myocardium can be diagnosed with certainty only if tubercle bacilli are demonstrated

c Actinomycosis usually occurs as an extension from the mediastinum or lung Masses of granulomatous tissue are customarily found in the pericardial sac although smaller lesions may only be seen microscopically The ray fungus is usually demonstrable

d Sarcoidosis a disease of unknown origin characterized by pseudotuberculous granulomata may involve the myocardium with typical interstitial or intrafascicular lesions

75 Parasitic Lesions of the Myocardium —a Although rare echinococcus cysts may be visible grossly They may be multiple in the septum or lodged between columnae carneae

b Cysticercosis (Taenia solium) is rare when present larvae may be found in the myocardium



Fig 30 An example of myocarditis frequently encountered in acute trichinosis Note the separation of the fibers (some of which exhibit degenerative changes) by an infiltration of eosinophiles and plasma cells (Courtesy of Armed Forces Institute of Pathology)

c. Infrequently myocardial granulomata may be encountered in schistosomiasis hematogenous dissemination of the ova may produce schistosomal tubercles of miliary size

d Trichinosis in severe cases may induce myocardial lesions It does not produce changes in the heart muscle that are visible to the naked eye While the larvae of the Trichinella spiralis almost never develop in the myocardium they may produce a lesion in which the heart is dilated and soft The muscle may appear pale brown and show areas of fatty change

Microscopic examination (Fig 30) may reveal loss of cross striations and sarcoplasm and various regressive nuclear changes in the muscle fibers In addition there may be diffuse infiltration by polymorphonuclear leucocytes and plasma cells and in some cases by eosinophiles. The parasites although usually indetectable may be demonstrated following digestion of a fresh specimen

e Trypanosoma cruzi (Chagas disease) —The protozoans lodge in the muscle producing necrosis and reactive inflammation in the interstitium (Fig 31)

f Plasmodium usually falciparum rarely vivax may in severe cases

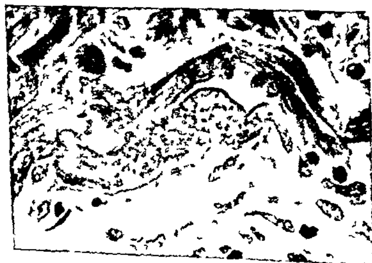


Fig 31 Myocarditis in trypanosomiasis cruzi (Chagas disease) The lesion shows collections of the leishman form of trypanosomes (having lost their undulating membranes and flagella) encysted in myocardial fibers (Courtesy of Armed Forces Institute of Pathology)

produce capillary thrombi with ischemic changes in the muscle. The parasites are visible with the agglutinated erythrocytes in the myocardial capillaries (Fig 32)



Fig 32 Myocardium in malaria (Falciparum) exhibiting slight edema, slight increase in interstitial cells (histiocytes, plasma cells and Anitschkow myocytes) with parasitized erythrocytes crowding capillaries and venules (Courtesy of Armed Forces Institute of Pathology)

g Toxoplasmosis may occur in the myocardium (Fig 33) producing focal necrosis associated with an eosinophilic and leucocytic infiltration

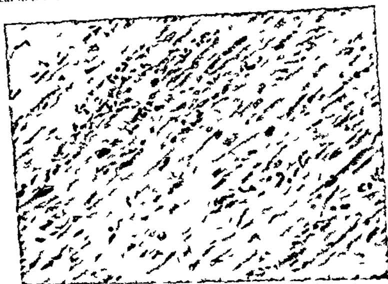


Fig 33 Acute focal myocarditis in toxoplasmosis exhibiting focal necrosis [illegible] cytes and polynuclear neutrophilic leukocytes Here and there an interstitial infiltrate of plasma cells may be seen (Courtesy of Armed Forces Institute of Pathology)

h Heterophyes ova in rare instances may be found in the valves and myocardium

i Sarcocystis a parasitic fungus infection of hogs and cattle may occur in man producing microscopic cysts in the myocardium

j Sarcosporidia common in lower animals may occur in man with demonstrable lodgment of the parasites in the myocardium

k Strongyloides larvae may be found in severe cases in the myo cardium

76 Hemochromatosis Myocardial involvement may be focal o diffuse With the latter moderate cardiac hypertrophy is usually found Microscopic lesions vary from moderate perinuclear deposits of hemo siderin to focal degenerative lesions with pigmentation and scarrin Macrophages laden with iron pigment are found in the microscop scars Calcification may occur in a focal pattern The muscle fibe may be normal in size or exhibit nuclear and muscle hypertroph

77 The Heart in Hyperthyroidism (thyrotoxic heart thyro heart hyperthyroid heart)—Some individuals dying with clinical e

dences of hyperthyroidism present slight or moderate enlargement of the heart without obvious cause Focal necroses diffuse degenerative changes similar to those in diphtheria variable types of cellular infiltration especially lymphocytic and patchy hyaline transformation are among the lesions which have been described These changes however are not specific

78 THE HEART IN MYXEDEMA (myxedema heart hypothyroid heart) —Because of inadequate necropsies the criteria for the diagnosis of myxedema heart are not established No special features either gross or microscopic have been recognized Hypertrophy with or without dilatation is usually described as well as hydropericardium Coronary arteriosclerosis of moderate or advanced degree is an almost constant finding The findings vary with the severity of myxedema Edema of the epicardial fat is common Interstitial edema and myxomatous change of the myocardium is less frequent Degeneration of muscle fibers and nuclear changes may be seen occasionally Extensive replacement fibrosis of the myocardium is not infrequent

79 THE HEART IN VITAMIN B DEFICIENCY (beriberi heart) — Changes in the heart in vitamin B deficiency diseases such as beriberi are rarely found at necropsy There may be both hypertrophy and dilatation particularly of the right side The pulmonary artery is markedly dilated and engorged as are the venae cavae

*Histologically* the myocardium may show no changes or the myocardial fibers may present various degrees of hydropic degeneration Longitudinal and cross striations may remain Interstitial edema congestion hemorrhage and swelling of the connective tissue may be present These changes however are not specific

80 NEOPLASMS OF THE MYOCARDIUM —Primary tumors are rare The benign tumors which may occur are fibroma angioma leiomyoma myxoma and rhabdomyoma The last is of congenital origin Metastatic carcinoma and sarcoma especially melanoma although rare are of interest as are the tumor or tumor like lesions of the anterior mediastinum or thymus that infiltrate the pericardium and penetrate the heart wall

81 INJURY OF THE MYOCARDIUM —*Blunt force injuries* of the heart are produced by crushing and contusion by tearing by bursting as a result of pressure from within by lacerations due to broken bones and by tearing of cardiac attachments

a *Crushing* injuries are produced by an extreme grade of violence

which shatters the chest wall and comminutes the cardiac muscle In children and adolescents the ribs are elastic and extreme compression of the chest may occur with few fractures

A severe force may fracture the sternum and ribs inflicting a *contusion* directly on the right ventricle or it may force the heart against the thoracic vertebrae bruising the left ventricle The violence producing cardiac contusion is usually lethal and the victim dies in a few minutes of associated injuries in other organs rarely of the cardiac injury itself The contusion in the myocardium appears as a dark red extravasation of blood of varying size and shape and also as a dark red hemorrhage in the adjacent epicardium *Microscopically* there are numerous red blood cells between the muscle fibers and in the subendocardial tissues

b *Tearing* of cardiac muscle may be caused by an extreme grade of trauma which crushes and exerts a tangential force on the body of the heart Less severe distortion of the heart may produce a complete transverse tear of one of the papillary muscles in the left ventricle It may cause a long thin shallow elliptical endocardial tear of the wall of the right atrium about a centimeter above the septal portion of the tricuspid valve and parallel to it The patient may live a few days with this latter injury and sometimes develops a small thrombus at the site of laceration *Microscopically* there is polymorphonuclear leucocytic reaction in the tissues around the thrombus Death in cases of this type is usually the result of other injuries

c *Bursting* ruptures of the heart are the result of a severe grade of violence applied to the chest which causes a rise of intracardiac pressure followed by perforation of the cardiac wall Most of these perforations are ovoid or stellate and of varying size involving the apex or mid portion of the right ventricle less frequently the apex of the left ventricle or the atria Sometimes ruptures are multiple Many of these injuries occur after falls from a height or in run-over cases and are associated with pericardial tears A slightly different rupture is produced by wood thrown by a circular saw rotating at high velocity which strikes the victim over the right costal cartilage area with severe impact One or two costal cartilages may be broken and a perforation of 1 to 2 millimeters in diameter may occur in the thin part of the ventricular wall between the pectinate muscles Death may occur almost immediately or after a slightly longer interval from intrapericardial hemorrhage A minor injury which occurs as a result of increased intracardiac pressure is a tear of a portion of one of the aortic cusps

d The heart may be *lacerated* by fracture of the sternum producing an ovoid tear in the middle of the right ventricle or by rib fractures which cause stellate lacerations analogous to the lacerations in the pericardium

e *Tearing of the cardiac attachments* follows severe injury to the chest wall which causes downward traction on the heart The aorta may be partially or completely torn transversely in the ascending portion but the other large vessels are not so frequently involved Cases occur in which all cardiac attachments are severed and the pericardium and diaphragm are torn so that the heart is ripped away and is loose in the chest or in the abdomen Sometimes the violence causes downward traction on the liver tearing the inferior vena cava inside the pericardial sac partially or completely in a transverse direction

The principle cause of death with these blunt force injuries of the heart is hemorrhage into the pericardial sac and cardiac tamponade if the pericardium is intact Hemorrhage occurs into one or both thoracic cavities if the pericardium is torn with death from loss of blood Other fatal complications are rare

*Penetrating wounds of the myocardium*—Penetrating wounds of the heart are produced by missiles and knives or other sharp pointed instruments It is necessary to locate a wound of entrance on the surface of the body in order to make a diagnosis of either a stab wound or bullet wound of the heart If the penetration injures the pericardium and myocardium without producing an opening which communicates with one or both pleural cavities hemorrhage into the pericardium occurs and death results from tamponade of the heart If the opening causes the pericardial sac to communicate with either pleural cavity, death is more gradual and is caused by loss of blood In some cases an acute suppurative pericarditis may be a fatal complication Rarely wounds of the left ventricle may not produce an immediate hemorrhage or infection but may give rise to a bland intracardiac thrombus on the endocardial surface Some wounds of the heart may heal and bullets or broken knife blades may be found in the myocardium enclosed in a fibrous capsule

82 FAT EMBOLISM is usually the result of injury to fat tissue especially fracture of the long bones Emboli may reach the heart on the arterial side of the circulation in large numbers after being forced through the pulmonary capillaries Streak like hemorrhages are found in the subendocardial layer or in the myocardium some of them having a small yellowish white center

*Histologically* fat emboli appear as ovoid globules in the inter

fascicular capillaries sometimes arranged in rows. The adjacent muscle fibers are necrotic and filled with globules of fat and red blood cells may surround the muscle fibers. The fat emboli and the degeneration in the myocardium may be demonstrated by osmic acid, sudan III or other fat stains (Fig. 34).



*Fig. 34. Fat emboli in the myocardium. Osmic acid stain. The fat emboli appear as black stained masses within small interfascicular capillaries.*

83. Air Embolism *may occur* after surgical operations, especially operations on the veins of the neck, or after injection of air into the uterus or other body *cavities*. *In some* cases death is sudden and air may distend the *right ventricle*, pulmonary arteries and the venae cavae. In other instances *the blood* may be frothy. Occasionally air reaches the left side of the heart through the pulmonary circulation and may enter the coronary arteries.

*The* diagnosis at necropsy depends on careful technique. *The* sternum is freed from the *mediastinum* from below upward as far as the second rib on each side, so that the veins at the base of the neck may not be cut. The pericardium is *flooded* with water and the heart is submerged in situ. Openings *are made* in both ventricles. If air is present, it bubbles through the water. Care must be taken to determine

if the gas inside the heart is air and not the gases of decomposition a determination which for the most part must be made from the history of the case and from the other findings at necropsy

84 POISONING may be accidental homicidal or suicidal Many poisons affect the heart as part of their general action on the body Potassium chlorate nitrobenzene or aniline form methemoglobin and the heart is tinged a dark purplish brown The prolonged action of phosphorus arsenic and other toxic substances produces diffuse fatty changes in the myocardium sometimes with small hemorrhages in the muscle and subendocardial layer of the septum of the left ventricle

*Carbon monoxide poisoning is either acute or delayed In the acute* cases the heart has a cherry red color due to carboxyhemoglobin In the delayed carbon monoxide deaths the victim survives the immediate effects of the poisoning but fails to recover from the results of the severe initial anoxemia The patient may die twenty four hours or more after exposure when all the carbon monoxide has been eliminated from the red blood cells and the blood and tissues have returned to their normal hue At necropsy lesions referable to anoxemia are found in the myocardium especially in the distal ends of the papillary muscles where the myocardial circulation is not as efficient as elsewhere (Figs 35 36) On longitudinal section of the papillary muscle

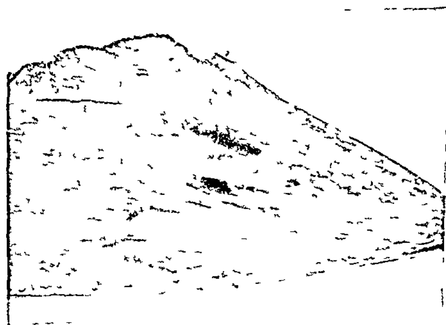


Fig [illegible] *Delayed carbon monoxide poisoning* Low power view of papillary muscle showing anoxemic necrosis and hemorrhage in muscle

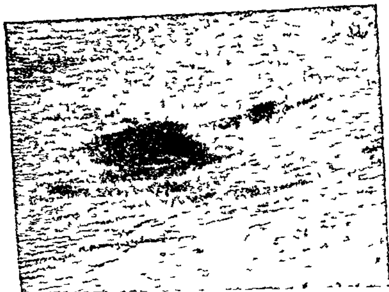


Fig. 56 Delayed carbon monoxide poisoning Higher power view showing acute necrosis with leucocytic infiltration and hemorrhage contrasted with normal myocardium

roughly circular dark red scattered hemorrhages about 1 millimeter in diameter may be present near the tip In other cases dark red and light red striations radiate out from the tendinous insertion into the myocardium forming a thistle shaped area about 15 millimeters in diameter

*Histologic examination* reveals that the smaller blood vessels are markedly distended and contain normal red blood cells laked erythrocytes or hyaline thrombi Sometimes hemorrhages may occur in between the muscle fibers and these fibers in places may be swollen eosinophilic without striations and with degenerated nuclei showing atrophy or necrosis polymorphonuclear leucocytes may infiltrate the edges of such foci In rare instances the myocardium may be generally permeated with hemorrhagic areas especially near the pericardium and endocardium histologically the lesions are as described above Because of the myocardial degeneration due to anoxemia mural thrombi may form in the left ventricle and may cause infarcts in some of the other organs

CHAPTER XI

# DISEASES OF THE CONDUCTION SYSTEM

## THE SINOATRIAL AND ATRIOVENTRICULAR NODES THE ATRIOVENTRICULAR BUNDLE (BUNDLE OF HIS) BUNDLE BRANCHES AND PURKINJE FIBERS

PHYSIOLOGIC evidence indicates that certain structures in the heart constitute a conduction system These are the sinoatrial node (S A node) the atrioventricular bundle (A V bundle bundle of His) the bundle branches and the Purkinje fibers With the exception of the S A node these structures are dissectable* (Figs 37 38)

*Histologically* the A V node bundle and bundle branches have certain differentiating characteristics common to all these structures and certain further characteristics pertaining to each part of the system All these structures are differentiated by (1) the endothelial lined sheath (2) the lesser number of myofibrils and (3) the increase in elastic tissue The node is further differentiated by its reticulated structure and the small size of its fibers The latter characteristic pertains to the bundle as well The left branch is characterized by the large size of its fibers (Purkinje fibers) in the distal four fifths and the sharp reticular component of the basement membrane The right bundle branch has no further differentiating characteristics The S A node is characterized by the small size of its fibers and the relatively large amount of collagenous fibers With advancing age all of these structures show an increase in collagen fibers in elastic tissue in fat and in density of reticulum

A qualitative method for the histopathologic study of the A V node bundle and bundle branches has been described ** For the quantitative determination of changes serial sections are required Routine study requires only hematoxylin eosin stain supplemented if necessary by elastic tissue–Van Gieson s stains (*Fig* 39)

There is not always a correlation between clinical disturbances in conduction and pathologic findings The reason for this is not known The following changes are described

* J W[illegible]dran and M Lev Ci[illegible] 4 863 19[illegible]1

** M Lev J Widran and E E Erickson A[illegible] *[illegible]es of Pathology* 5[illegible] 3[illegible]19[illegible]

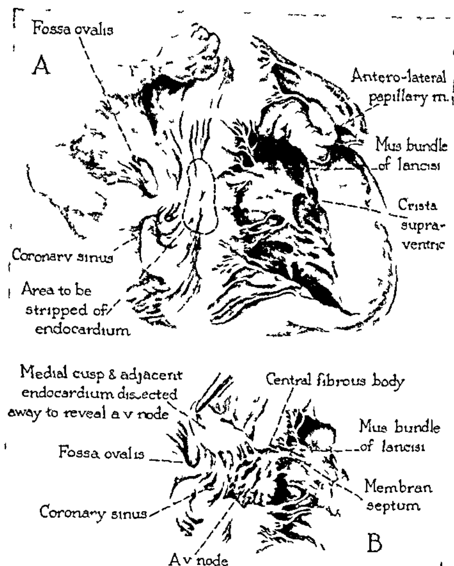



FIG 37 A Drawing of the opened right heart to show landmarks and site for dissection of the A V node bundle and bundle branches B The exposed A V node after dissection

85 DEGENERATIVE CHANGES are the most frequent lesions found The right branch and the anterior subdivision of the left branch are chiefly affected because they have a common blood supply

The underlying lesions leading to degeneration of the conduction tissue are (1) recent or old infarcts of the interventricular septum (2) severe coronary sclerosis and (3) pressure from expanding lesions such as calcific deposits especially in the interventricular septum neoplasms and gummata

a Changes secondary to vascular disease are those resulting from reduction or interruption of blood supply The end result in either case is fibrosis or calcification The vessel most often affected is the anterior descending ramus of the left coronary artery The conduction system may show advanced fibrosis without other myocardial change and without occlusion of an artery

b Changes secondary to pressure result from expanding lesions such as calcific deposits gummata and neoplasms The calcium mass is frequently located at the base of the anterior leaflet of the mitral valve

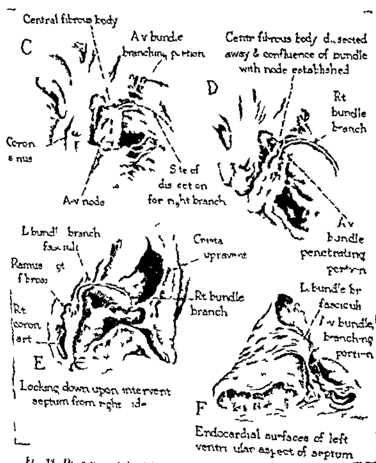



Fig 38 Dissection of the A V node bundle and bundle branches continued from figure 3
C The A V bundle branching portion dissected
D A V bundle penetrating portion and right bundle branch dissected
E Beginning of left bundle dissected
F Further dissection of left bundle branch

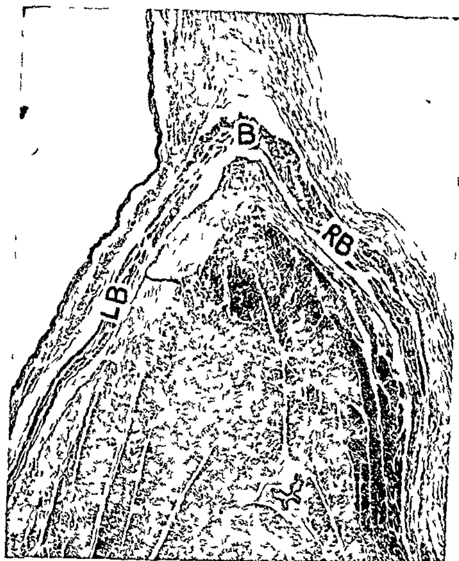



FIG. 39. Photomicrograph showing A-V bundle and right and left bundle branches (hematoxylin-eosin stain ×20). B indicates bundle; RB, right bundle branch; LB, left bundle branch.

Note the lobulated space through which the bundle branches course. (Courtesy of M. Lev, J. Widran and E. E. Erickson.)

or in the aortic valve and may extend into the interventricular septum at the junction of the membranous and muscular portions. Gummata and neoplasms likewise may be found in approximately the same site.

*Histologically* the changes in the tissue depend upon the underlying lesions. Thus in a myocardial infarct there may be widespread destruction of the bundle tissue. In expanding lesions the conduction tissue may be incorporated in the lesion itself or affected by radiating scar

tissue Degeneration may occur when any adjacent expanding lesion exerts pressure on the conduction tissue Above and below the site of the lesion the bundle and its branches may be normal

86 INFLAMMATORY LESIONS —a *Rheumatic fever* —Lesions in the conduction system are frequently encountered in the acute phase of this disease *Histologically* in active cases there may be an accumulation of lymphocytes with a few plasma cells and histiocytes Sometimes there may be numbers of polymorphonuclear leucocytes Edema is occasionally seen and the amount of elastic tissue is frequently increased particularly in active cases of long duration Rarely Aschoff bodies (Fig 40) may be demonstrated in the bundle

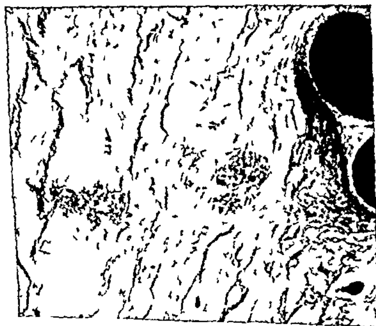


Fig 40 Cross section of horizontal conduction system from a case of active rheumatic fever This low power photograph shows Aschoff bodies in edematous and inflamed bundle tissue the large blood vessels were injected

The vascular lesions are similar to those occurring in the coronary arteries in rheumatic fever (see Chapter V) In the majority the intima exhibits proliferation without elastic changes followed by musculoelastic hyperplastic changes Many cases show dilated capillaries some of which are thickened Vascular changes are more conspicuous and frequent than exudative ones A frequent form of inflammation occurs in

the collagenous extension of the septum fibrosum A distinct increase in vascularity as well as in inflammatory cells takes place These lesions may be contiguous to those in the root of the aorta or the ring of the aortic valve In inactive cases fibrotic lesions may occur

b *Syphilis* involves the conduction system in the form of (1) a gumma located in the interventricular system (2) diffuse gummatous myocarditis or (3) a scar The continuity of the conduction system may be interrupted by the gummatous mass

c In *bacterial endocarditis* both acute and subacute there may be extension of the inflammatory process from the valves usually the aortic into the ventricular septum Lymphocytes and a few polymorphonuclear leucocytes are found together with destruction and necrosis of the area affected Healing with subsequent calcification may take place

d In *diphtheria* the conduction system may be invaded by lymphocytes and eosinophiles These collections tend to be perivascular and may be so extensive as to disrupt the continuity of the bundle The *muscle is edematous and where not replaced by cellular elements* is swollen granular and stains poorly

87 Congenital Lesions —Although congenital heart block partial or complete has been frequently reported clinically few cases have been studied adequately at necropsy Where this has been done absence of or disruption in continuity or extensive fibrosis of the conduction system has been found Lesions have been reported associated with aortic stenosis and atresia and with defects of the ventricular septum In the latter case however the bundle may be uninterrupted as it follows a circuitous course around the periphery of the defect

88 Neoplasms when primary cause heart block more often than metastatic lesions Together they make up a small fraction of the cases Atrial angiomata and lymphangio *endotheliomata are the types of* primary lesions which have been described

CHAPTER VII

# DISEASES OF THE ENDOCARDIUM AND VALVES

ENDOCARDITIS refers to the inflammatory changes of the lining of the valves and cardiac chambers which may be the result of either a bacterial agent or a systemic disease (rheumatic fever systemic lupus erythematosus) or due to factors yet unknown

The diagnosis of *bacterial endocarditis* should include the causative bacterial agent In bacterial endocarditis micro-organisms can usually be found in spreads from vegetations on the valves or mural endocardium The diagnosis should include antemortem and postmortem bacteriological as well as histological findings

89 ACUTE BACTERIAL ENDOCARDITIS (infective vegetative ulcerative) —The bacteria usually responsible are the hemolytic streptococci pneumococci and Staphylococcus aureus The gonococcus meningococcus Bacillus pyocyaneus Friedlander's bacillus *Spirillum minus* and Escherichia coli are occasionally found

Vegetations are generally large and exuberant but may be small and flat occasionally inconspicuous and difficult to recognize especially if located on the ventricular aspect of the atrioventricular valves The vegetations are usually gray sometimes green or red soft and friable The vegetations are most often superimposed upon a previously diseased or congenitally deformed valve although some may occur on a normal valve The left side of the heart is more frequently involved than the right Ulceration is a common feature as are also *aneurysms* of the leaflets and of the sinuses of Valsalva Perforation of the leaflets or of the interventricular septum and other segments of the heart wall and destruction of the chordae tendineae may occur Destructive features are much more conspicuous in the acute than in the subacute form

*Microscopically* (Fig 41) the vegetations consist of fibrin enmeshing fused erythrocytes many bacteria often in colonies blood platelets and polymorphonuclear leucocytes Fragments of valve substance showing varying degrees of necrosis are often incorporated and may be seen with special stains Large elongated mononuclear cells may be seen arranged at right angles to the injured surface forming a basal



Fig 11 Acute bacterial endocarditis of mitral valve due to hemolytic *streptococcus* The swollen leaflet is necrotic in place and is heavily infiltrated by fragmented polymorphonuclear leucocytes The endothelial surfaces are ulcerated and partly covered by surface growth of darkly staining bacterial colonies

palisade There is often purulent infiltration at the base of the lesion with widespread necrosis and ulceration Healing in untreated cases is generally minimal but may be seen in the base of the vegetations In cases that have received antibiotics there is a distinct tendency to partial healing with active proliferation of mononuclear cells fibroblasts hyalinized connective tissue and even foci of calcification Complete healing may occur (see paragraph on *Healed stage of Subacute Bacterial Endocarditis*)

90 Subacute Bacterial Endocarditis (endocarditis lenta subacute or chronic infective endocarditis) —a *Active stage* (with bacteremia) — A non hemolytic streptococcus (Streptococcus viridans) is the most common etiological agent Other organisms include enterococcus gonococcus Hemophilus influenzae pneumococcus Brucella group diphtheroids and the anthrax bacillus Higher bacteria yeasts or fungi Actinomyces bovis Histoplasma capsulatum and Streptobacillus moniliformis have been reported Mixed infections occasionally coexist

Valvular deformities due to rheumatic fever are frequent precursors An active rheumatic lesion is often present simultaneously especially in younger persons Congenital anomalies particularly bicuspid aortic

valve and sclerotic valves including occasionally those due to syphilis may pre exist

The vegetations are mainly located on the valve surfaces but may be present on the parietal endocardium especially that of the left atrium and the upper portion of the left ventricular aspect of the interventricular septum The left side of the heart is preponderantly involved The vegetations may extend on and even ulcerate through the interventricular septum or spread along the chordae tendineae and papillary muscles The vegetations may be discrete or confluent flat and granular or occasionally very large polypoid green or yellow friable or firm The sinuses of Valsalva are rarely penetrated and followed by pericarditis Pericarditis associated with uremia or that due to rheumatic fever may be present

*Microscopically* (Figs 12 13) the vegetations consist principally of platelet thrombi containing colonies of bacteria on and near the surface usually covered by a sheet of fibrin containing few polymorphonuclear leucocytes and red blood cells Large multinucleated cells frequently vacuolated or elongated mononuclear cells at right angles to the surface may occur at the base and infiltrate the substance of the valves These cells may contain large numbers of bacteria Remnants of valve substance in varying stages of necrosis are frequently noted Granulation tissue may occur at various levels beneath the vegetations There may be partial or complete organization with or without calcification or occasionally even bone formation As in acute bacterial endocarditis but less frequently the chordae tendineae may rupture and the valve substance may undergo aneurysm formation or perforation

b *Healed stage* (bacteria free) —Since the advent of antibiotic therapy complete healing is encountered more frequently than previously described The so-called bacteria free stage in which the vegetations disclose evidence of healing and in which the organisms are either absent or dead in no way differs from the lesion in which the bacterial activity has been reduced or completely destroyed by antibiotics The vegetations undergo fibrosis hyalinization and occasionally calcification forming irregularly nodular masses with ensuing deformity which may produce stenosis or insufficiency of the valve In the partially healed stage granulation tissue may be prominent but a few bacteria persist In the completely healed stage fibrosis with hyalinization and calcification are noted simulating the changes incident to chronic rheumatic valvulitis In some instances particularly treated cases there may be only minimal thickening of the cusps with rupture of the

chordae tendineae and perforation of the leaflets No bacteria ar
demonstrable in this stage

The focal type of glomerulonephritis with fibrosis and hyalinizatio
(healed embolic ) may be noted and help identify the nature of th

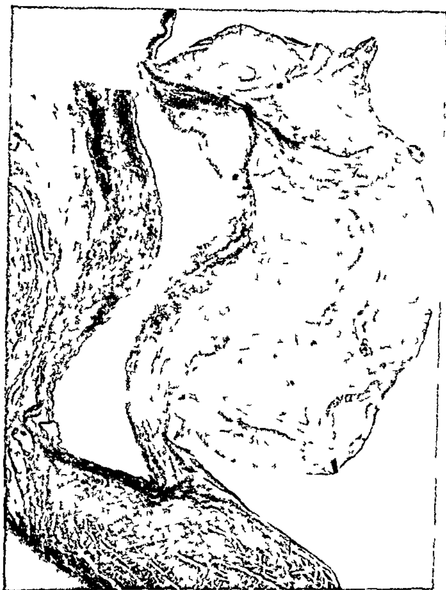


FIG 42 Subacute bacterial endocarditis of aortic valve The ventricular aspect of leaflet is covered by a bulky vegetation which merges rather imperceptibly with the remains of the valve substance It is composed largely of fibrin platelets and amorphous debris deposited on an exuberant growth of granulation tissue The elastic layers of the leaflets are reduplicated and near the base there is evidence of old fibrous proliferation



Fig 43 Subacute bacterial endocarditis The ulcerated surfaces are covered by fibrin platelets and masses of bacteria The substance of the leaflet is heavily infiltrated by large hyperchromatic mononuclear cells.

valvular lesion Generally acute bacterial endocarditis is readily differentiated from the subacute variety in view of the more extensive ulceration necrosis and absence of evidence of repair in the former The identification of the lesion may be difficult in those instances that have received antibiotics for fibrosis and hyalinization are common to both forms

91 Indeterminate Endocarditis includes verrucous or thrombotic endocardial lesions characterized by thrombotic deposits of blood elements They are independent of direct bacterial infection and bear no relationship to rheumatic fever

a *Atypical verrucous endocarditis* (Libman Sacks) is a pathological entity occurring frequently in systemic lupus erythematosus The gross differentiation of this form of endocarditis from that due to bacterial infection and rheumatic fever is generally simple because of the form and size of the verrucae (Fig 44) There is frequent involvement of the ventricular aspect of the valve cusps in particular the endocardium of

chordae tendineae and perforation of the leaflets No bacteria are demonstrable in this stage

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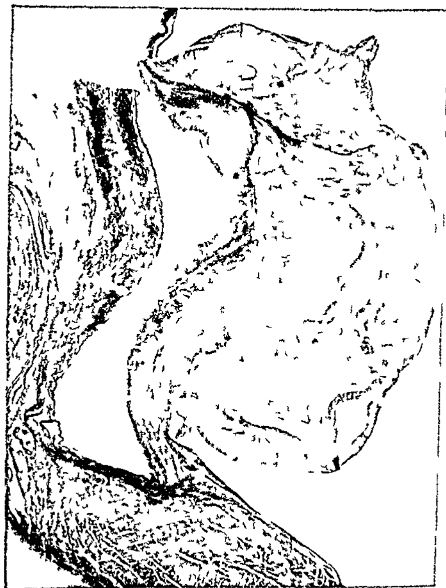


Fig 12 Subacute bacterial endocarditis of aortic valve The ventricular aspect of leaflet is covered by a bulky vegetation which merges rather imperceptibly with the remains of the valve substance It is composed largely of fibrin platelets and amorphous debris deposited on an exuberant growth of granulation tissue The elastic layers of the leaflets are reduplicated and near the base there is *evidence of old fibrous proliferation*

this group is not clear despite its relatively frequent occurrence Since these lesions are seen commonly in infectious or chronic wasting diseases it has been assumed that the products of bacteria or toxins in chronic disease may be instrumental in their production Micro-organisms are not responsible for their formation but may be secondarily implanted




Fig 45 Atypical verrucous endocarditis of mitral valve The shortened thickened and eroded leaflet has a small verrucous protrusion on its atrial aspect (A) and a larger mass of granular vegetative material (B) on its ventricular aspect caught between the leaflet and the wall of the left ventricle These deposits are free from bacteria

In its verrucous form the lesions are firm gray pale yellow or blood stained flat or raised *nodules or plaque like* lesions along the line of closure of the mitral valve and occasionally of the aortic valve either continuous or discrete The smaller lesions may simulate rheumatic endocarditis However they are *often* larger than rheumatic verrucae They vary in size from pinhead *to* pea size or even larger and occa

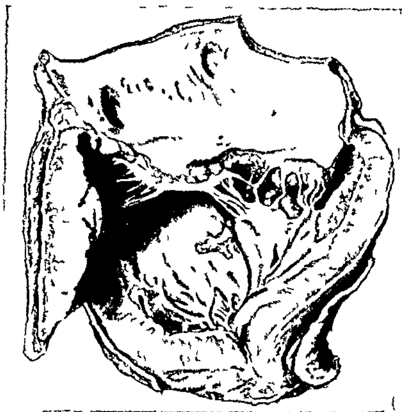


FIG 44 Atypical verrucous endocarditis of mitral valve Vegetations extend along both aspects of the valve in the pocket and on the wall of the left ventricle

the valve pockets with flat extensions of finely granular verrucae upon the mural endocardium

*Microscopic examination* reveals hematoxylin bodies as the distinctive feature The verrucae consist of granular masses in part fibrin in the meshes of which are red and white blood cells (Fig 45) The most significant features however are groups of ovoid hematoxylin stained bodies of the size of fibroblastic nuclei or even smaller Similar bodies are found singly throughout the verrucae At the base of the verrucae there is infiltration with macrophages lymphocytes many fibroblasts and plasma cells Between the collagen fibers there is frequently a deposit of clumpy or fibrillar fibrinoid material Occasionally vascular emboli are found in the myocardium Bacteria are not demonstrable ordinarily but bacterial implants may be superimposed

b *Thromboendocarditis* (including non rheumatic verrucous endocarditis non bacterial thrombotic endocarditis endocarditis simplex toxic endocarditis and terminal endocarditis) —The pathogenesis of

leaflets may be otherwise normal the lesion being thus distinguishable from rheumatic verrucae

*Histological examination* is essential for this differentiation The larger discrete thrombotic types also show a notable absence of underlying inflammatory change The vegetation may consist of fresh blood elements with lamination of fibrin as a prominent feature Although examination of the myocardium discloses no Aschoff bodies the thrombotic deposits may be superimposed upon chronic rheumatic valvulitis

Occasionally in older individuals tag like processes may be seen on the *noduli arantii* of the aortic cusps These may simulate healed endocardial vegetations However histologically they consist of hyalinized or edematous myxomatous connective tissue without thrombosis or any evidence of reactive inflammation They are considered to be degenerative endocardial lesions

92 TUBERCULOUS ENDOCARDITIS is rare and when found usually occurs in instances of miliary tuberculosis

93 SYPHILITIC ENDOCARDITIS is rarely primary and results rather from contiguous involvement of the endocardium by way of the aortic lesion (see Chapter III)

94 ANNULAR SCLEROSIS is a fibrocalcific deposit within the valve ring and is common in senescence It may involve either the mitral or aortic rings or both producing a rigid collar like annulus The free parts of the valve may remain normal or the leaflets may present sclerotic foci of thickening Valve function may or may not be disturbed *Microscopically (Fig 17)* the principal features are cholesterol deposits elastic tissue proliferation fibrosis and hyalinization frequently with calcification of the rings Ossification may occur Occasionally a calcified and ossified ring may produce pressure ulceration of the base of the valve

95 VALVULAR ATHEROMA (*endocardial atheroma*) is probably an early stage of endocardial or valvular sclerosis which frequently involves the mitral valve Typical lesions are seen on the ventricular aspect of the anterior mitral leaflet and consist of soft irregular broad or linear yellow plaques involving the endocardium which do not produce valvular deformity *Microscopically* localized collections of lipid material are noted Fibrosis and local calcification may occur

sionally may form a massive discrete polypoid or even pedunculated lesion The underlying valves may show only minimal or no evidence of previous damage The mitral and aortic valves are most commonly affected Neither the mural endocardium nor the chordae tendineae are involved Only the larger lesions may produce valvular deformity

Thrombotic deposits also may be implanted on a calcified or eroded leaflet or over mural endocardial plaques in the various chambers Microscopically the verrucous excrescences (Fig 16) consist of compact



Fig 16 Thromboendocarditis The endothelial surface of a scarred and deformed valve is covered by a fresh deposit of deeply staining fibrin Signs of inflammation in the region of the thrombus are lacking

blood elements chiefly fibrin blood platelets form cells resting upon edematous smudgy eosinophilic connective tissue There are no bacteria present nor is there any reactive inflammatory lesion adjacent to the thrombotic mass Occasionally however in the older lesions connective tissue proliferation may be noted with fibroblasts and histiocytes while the healed lesion consists of a fibrous nodule or irregular collagenous thickening near the free edge of the valve The valve

nodules in their substance rather than in the free margins In advanced cases calcific nodules exaggerate the deformities of the cusps The deposits may be rounded or ragged and frequently penetrate the endothelial layers especially along the ventricular surfaces The margins of the aortic cusps may be sharply defined or thickened Calcific masses may be located within the sinuses of Valsalva producing nodular excrescences on the valve surfaces and may occasionally even fill a sinus of Valsalva There may be fusion at the commissures of the valve cusps The deformity may have the appearance of a bicuspid valve

*Histologically* (Fig 48) changes are seen at the base of the cusp in the form of areas of fibrosis and hyalinization Deposits of lipid and



Fig 48 Calcification of the aortic valve A large mass of calcified material is imbedded in fibrous tissue which protrudes into the sinus of Valsalva The proximal portions of the leaflet and the entire ventricular aspect show little change



FIG 17 Calcification of the annulus of the mitral valve A large mass of calcium which has become fragmented in preparation is situated at the base of the valve and extends into the ventricular myocardium The valve leaflet is shortened and thickened

96 VALVULAR SCLEROSIS (calcific nodular valvular sclerosis calcareous valvular disease Monckeberg's aortic sclerosis primary ascending sclerosis of the aortic valve) —In older non rheumatic individuals the aortic cusps may be the seat of primary degenerative changes which produce rigidity and deformity of the valves accompanied by stenosis of the orifice with or without insufficiency Since sclerosis and calcification of the aortic valve are often a sequel of aortic valvulitis it may be impossible *occasionally to differentiate primary aortic valve sclerosis* from an old calcific rheumatic aortic valve In the primary form the fibrosis and calcification begin in the arterial surface of the aortic valves extending up the leaflets or to the sinuses of Valsalva

The degree of deformity of the aortic valve determines the presence of left *ventricular* hypertrophy The ascending aorta may appear normal In some cases the aortic cusps are thickened by small fibrotic

adherent and leave a rough surface when detached Histologic study of the site of attachment is necessary in doubtful cases and will reveal the presence and nature of underlying tissue changes Note should be made of the presence of platelet columns fibrin leucocytes red blood cells and evidences of organization When inflammatory changes occur special stains to identify micro-organisms should be employed

100 ENDOCARDIAL BLOOD CYSTS (so-called blood cysts) are seen rather frequently in newborn infants occasionally in childhood and rarely in adults These are small circumscribed nodular dark red cyst like lesions usually on the atrial surface of the mitral and tricuspid valves They vary from pinpoint to 1 millimeter in diameter and may be multiple Histologically they appear as simple or multilocular endothelial lined spaces filled with red cells and give the appearance of blood lacunae The adjacent connective tissue is moderately cellular

101 ANATOMICAL SIGNS OF VALVULAR DYSFUNCTION —The main disturbances of function resulting from deformed valves are stenosis or insufficiency or both Although rheumatic fever is the most frequent cause inflammatory and degenerative lesions such as syphilis sclerosis of valves including the Monckeberg type bacterial endocarditis either active or healed and congenital anomalies may also produce deformities with resulting valve dysfunction

a *Insufficiency (regurgitation incompetency) of a valve* is present when complete closure during life was impossible and the valve orifice is enlarged The diagnosis can be made when the altered valves cannot be made to coaptate Fibrosis and retraction of the semilunar cusps or widening of their commissures or flattening of the lateral portions of the cusps due to their adherence to the adjacent intima of the aorta or pulmonary artery or fusion and retraction of the chordae tendineae with dilatation of the rings of the atrioventricular valves produce insufficiency

Collateral evidences are noted mainly in advanced degrees of valvular dysfunction and consist of

(1) Hypertrophy and dilatation of the chamber behind the incompetent valve

(2) Mural endocardial pockets often with the openings of the pockets directed toward the incompetent valve orifice or patchy fibrosis in similar locations (Fig 49)

(3) Perforation of valve leaflets resulting from disease or trauma

b *Stenosis of the orifice of a valve* is present when fibrosis of a valve

calcium occur and coalesce to form large irregular nodules Occasionally there is metaplasia to bone A mild non specific cellular reaction may be present

97 Mural Endocardial Fibrosis—Various forms of mural endocardial thickening have been described in infants as well as in adults The pathogenetic factors leading to this often conspicuous and diffuse endocardial fibrosis are still unknown In the adult cardiac hypertrophy *and dilatation with mural thrombi may be conspicuous features*

98 Rheumatoid Valvulitis—In some patients with rheumatoid arthritis microscopic lesions resembling the subcutaneous nodule may be found in the valvular cusps

99 Mural Thrombosis (*endocardial thrombosis*) *may result from* (1) inflammation injury necrosis and fibrosis and (2) slowing and abnormal eddying of the blood stream Thrombi may form on the endocardium of any of the cardiac chambers including the auricular appendages

The marantic thrombus (stasis thrombus) is frequently found in the auricular appendages most often in atrial fibrillation and congestive heart failure Ball or globoid thrombi occasionally lie free in the atrial chambers or may be attached to the endocardium by a thin pedicle These may act as ball valve obstructions by protruding into the atrioventricular orifices They are most often found in association with stenosis of the mitral or tricuspid valves Atrial mural thrombosis is encountered in rheumatic heart disease with severe mitral stenosis and rheumatic endocarditis and may serve as the nidus for bacterial implantation

The majority of the mural thrombi in the ventricles form upon damaged endothelium overlying an area of myocardial infarction which has extended to the endocardium With extensive infarction of the interventricular septum mural thrombosis may also develop in the right ventricle Mural thrombi however are found predominantly in the left ventricle and may be small or large enough to fill a third or more of the chamber Mural thrombosis may occur in ventricular aneurysms incident to myocardial infarction The large thrombi are attached at their bases are friable and laminated and may be organized On section they vary in color but are predominantly red or gray They may be the source of systematic or pulmonary emboli

It is necessary to distinguish between thrombi and postmortem clots Aside from structural differences clots are easily removed thrombi are

tract of the left ventricle below an incompetent or stenotic aortic valve Their etiology is not clear Their presence suggests valvular insufficiency They vary in number and may be discrete or conglomerate Some, known as diastolic pockets may have openings a few millimeters wide directed toward an incompetent aortic valve orifice Others may occur in the left atrium just proximal to the base of the mitral valve and facing the apex *Histologically* the lesions consist of thickened fibrous mural endocardium

has produced fusion and stiffening of the cusps with narrowing of the valve orifice

Hypertrophy and dilatation of the chamber behind the valvular deformity usually occur and are roughly proportional to the degree of stenosis Mild grades of stenosis without such secondary changes are seen Advanced valvular stenosis is commonly accompanied by insufficiency Additional evidence may be circumscribed endocardial thickenings or pocket formation beneath the affected orifice

No satisfactory method of measuring valvular orifices has been established Tables which give the average normal circumference of the various orifices in the adult heart are unreliable *e g* aortic 7 to 8 centimeters mitral 9 to 11 centimeters tricuspid 11 to 13 centimeters Measuring orifices by the insertion of the finger or by a graduated wooden cone may dislodge vegetations The diagnosis of stenosis or insufficiency should rest on the anatomical factors described

c *Endocardial pockets* are mural endocardial thickenings (Fig 19) The most common are semilunar pockets on the wall of the outflow



Fig 19 Endocardial pocket of left ventricle in aortic stenosis and insufficiency

tract of the left ventricle below an incompetent or stenotic aortic valve Their etiology is not clear Their presence suggests valvular insufficiency They vary in number and may be discrete or conglomerate Some, known as diastolic pockets may have openings a few millimeters wide directed toward an incompetent aortic valve orifice Others may occur in the left atrium just proximal to the base of the mitral valve and facing the apex *Histologically* the lesions consist of thickened fibrous mural endocardium

CHAPTER VIII

# DISEASES OF THE AORTA AND PULMONARY ARTERY

102 ACUTE AORTITIS* may occur in any general infection such as pneumonia meningitis scarlet fever septicemia and in acute bacteremias with or without bacterial endocarditis The lesions may be difficult to detect grossly They vary with the age severity and type of infection When associated with bacterial endocarditis there may be vegetations or ulcerations in the intima

*Histologically* not only the intima and subintima but sometimes the deeper coats may exhibit an inflammatory response varying from a mild to a frankly suppurative reaction Special stains may reveal bacteria

103 GIANT CELL MESAORTITIS — A rare histologic lesion of the aorta of unknown etiology The lesion consists of patchy destruction of the elastic lamellae involving the middle third of the media with secondary inflammatory changes The most striking feature of the latter is the presence of multinucleated giant cells The lesion may be distinguished from syphilis by the absence of granulation tissue and adventitial changes and by negative serological evidence

104 ARTERIOSCLEROSIS OF THE AORTA OR PULMONARY ARTERY refers to a variety of lesions of unknown etiology manifested by regressive and proliferative changes in the intima with or without secondary changes in the media A wide variety of deposits may be found singly or in combination in individual lesions including lipids calcium soaps and phosphates and intercellular substances such as elastic fibers reticulum collagen bone and blood elements The chief organic effects are loss of elasticity dilatation and deformities Intimal lipid deposits atheroma atherosclerosis atherosclerosis with calcification and erosion and intimal sclerosis are included under this term The use of the term *Atheroma* is restricted to focal subendothelial lesions consisting of small accumulations of debris and extra-cellular lipid *Atherosclerosis* refers to atheromatous lesions accompanied by proliferation of the

* For discussion of Rheumatic and Syphilitic Aortitis see Chapters II and III respectively

intima *Intimal sclerosis* refers to foci of hyperplasia which are apt to occur in hypertension and are not primarily the seat of lipid deposition

*Arteriosclerosis* not infrequently affects the *pulmonary artery and its branches* It may be due to the increased pulmonary artery pressure found in cases of mitral stenosis and in any pulmonary disease of long standing *e g* pneumoconiosis and bronchiectasis and pulmonary conditions secondary to kyphosis and other deformities of the thorax The lesions as a rule are not far advanced and are practically never accompanied by erosion or calcification although they may furnish a site for thrombosis Primary pulmonary arteriosclerosis is rare

The early atheromatous lesions consist of small round gray or yellow areas a few millimeters in diameter They usually project slightly on the inner surface of the aorta Later they tend to fuse as large round or irregular soft yellow plaques The surface of the older plaques may become pale blue and *translucent* The superficial intima may slough leaving a ragged atheromatous erosion which is sometimes covered by thrombi Calcareous deposits may occur in the atheromatous areas and may project through the intima

Intimal sclerosis is like the atheromatous variety in distribution and is most marked around the mouths of small branches notably those of the intercostal arteries The lesions may be pale blue and translucent raised or retracted and wrinkled depending upon the state of the underlying media Occasionally it may be superimposed on syphilitic aortitis

The most advanced lesions of arteriosclerosis are most often found in the lower abdominal aorta The thoracic aorta is less severely and less often affected Therefore the coronary orifices are not as frequently encroached upon as in syphilitic aortitis The aorta may be dilated and thin due to impairment of the media especially in elderly individuals *Elongation and tortuosity of the vessel* are rather frequently seen in the advanced cases Aneurysm in uncomplicated arteriosclerosis of the aorta is infrequent However the abdominal aorta since it undergoes the most marked changes is occasionally the site of aneurysms especially in the aged

*Histologically* in the early stages of atheroma intra and extracellular lipids are demonstrable in the thickened intima The lipid may be contained in subendothelial fibroblasts and foamy macrophages even before the formation of the atheromatous plaque Later new formation or splitting of elastic fibers with intimal sclerosis may become prominent The muscle fibers of the media may atrophy in marked cases and may or may not be replaced by fibrous tissue Elastic fibers

lose their wavy appearance and lie close together In more advanced cases the lipid is usually visible as a disc shaped mass chiefly extra cellular sometimes mixed with blood elements Lipid filled macrophages cholesterin or fatty acid crystals granular plasma shreds of fibrin and even red blood cells may be demonstrable within the plaques Still later hyalinization of the intimal connective tissue is conspicuous Sometimes it is thinned out to a narrow strand or may even be ulcerated Calcium is frequently precipitated in these lesions in broad bands flat plates or as fine amorphous granules Hemorrhage into the plaque together with fibrin deposition or thrombosis may occur in the superficial zones An ingrowth of new blood vessels chiefly capillaries may be found in the underlying wall In advanced cases focal collections of lymphocytes are not uncommon in the adjacent tissue

103 CALCIFICATION OF THE AORTA —a *Medial calcification* —Medial deposits may occur in aortas exhibiting no other changes It is most frequently found in older individuals The aorta may exhibit dilatation with slight intimal thickening and atrophy of the muscle fibers There is stretching and separation of the elastic fibers Fixative containing acid or highly acid preparations of hematoxylin will remove such deposits The lesion is microscopic (Fig 50) and consists of basophilic, finely granular material dispersed in the middle third of the media The deposits vary in amount in different areas They occur independently of other degenerative lesions and bear no relationship to any known inflammation

b *Subintimal (metastatic) calcification* is a rare form of arterial disease that may involve the aorta and is characterized by calcification in the subintimal zone in close relation to the internal elastic lamina It occurs in hypercalcemic states and should be looked for at necropsy in persons with hyperparathyroidism osteitis fibrosa cystica osteomalacia and other osteoporotic diseases It has been encountered in chronic myelogenous leukemia with severe osteoporosis and in chronic nephritis associated with bone resorption especially in children ( renal rickets ) The intima may appear intact grossly or it may present transverse or circumferential ridges at sites of calcification sometimes covered by small thrombi

*Histologically* basophilic amorphous or plate like deposits of calcific material may be seen about the elastic fibers of the musculo-elastic layer even impregnating ruptured fibrils If the deposits have been present for some time fibroblastic proliferation may be marked about them and even metaplastic bone formation may occur This type of

calcification should be differentiated from medial calcification inasmuch as the media may show no changes

106 MEDIAL NECROSIS (medial degeneration medionecrosis aortae idiopathica cystica) is a degenerative process involving the aorta characterized by degeneration of the muscle and elastic fibers It may be recognized on gross examination

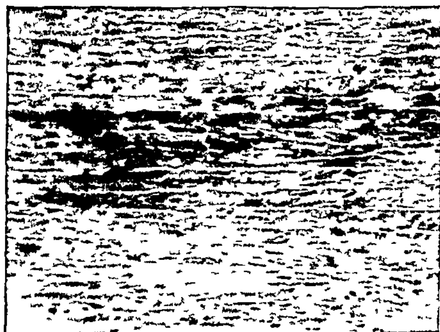


FIG 50 Medial calcification of the aorta Fine granules of calcium precipitate are deposited in the middle third of the media Where these are most abundant the tissue appears dark staining The unstained elastic fibers can be seen to course through the calcified portion and are themselves not involved

Features that may be of assistance are

a Spontaneous rupture of an aorta with an apparently normal intima

b Dissecting aneurysm of non traumatic origin not obviously associated with other disease

c Local defects in the media with thinning of the muscle layer covered by a somewhat wrinkled intima These lesions may be confused with foci of syphilitic mesaortitis

d Tendency for the wall on section to split into two layers usually in the media

e Presence of cystic spaces in the media

The essential *microscopic lesion* (Fig 51) is disappearance of smooth muscle cells from the middle third of the media as reflected by depletion of nuclei in this area The affected areas vary in extent and are never sharply defined There may be no evidence of current necrosis or of inflammation Often there are small cysts containing a basophilic mucoid substance roughly oval in shape with their long axis parallel to the surface There may also be irregular crevices These may be

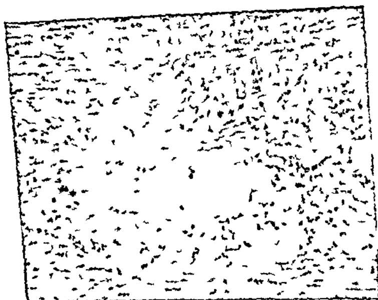


Fig 51 Medial degeneration of the aorta Here is an irregular gap in the media filled with mucoid substance and markedly depleted of nuclei The cells at the margins of this lesion are irregularly arranged without any inflammatory cellular infiltration

artefacts but indicate nevertheless that the tissues are but loosely held together The presence of blood elements in superficial crevices indicates antemortem tearing The other coats show no change Zones of healing may be encountered in which the new muscle is laid down in small whorls or at right angles to the longitudinal bundles

Combined elastic tissue and van Gieson s stains are helpful in establishing the diagnosis There is loss of muscle tissue and a relative increase of connective tissue In the areas of nuclear losses there is rarefaction of the elastica due either to diminution in the number and thickness of its fibers or to their displacement and concentration at the margins of the lesions

107 ANEURYSM — An aneurysm particularly of the ascending aorta

due to disease of the wall is to be distinguished from dilatation due to hypertension senility or similar conditions In the pulmonary artery or its main branches aneurysms occur rarely except in small branches traversing the walls of tuberculous cavities Syphilis plays the most important role in the development of aneurysm of the aorta (Chapter II) Other less frequent causes are arteriosclerosis acute infections inflammations and injuries

Aneurysms of the aorta may be due to syphilis or arteriosclerosis mycotic (embolic) or traumatic They may be located in the sinuses of Valsalva or any portion of the aorta They may be single or multiple The etiology can usually be determined Saccular and fusiform aneurysms are the two structural varieties The saccular is most commonly found in the ascending thoracic aorta Aneurysms of the abdominal aorta are usually caused by arteriosclerosis and are uncommon except in the aged Multiple aneurysms are usually saccular The tissues in the immediate neighborhood are compressed Contiguous structures may be eroded or displaced Rupture and hemorrhage occur in approximately half the cases Rupture may occur into the pericardium pleura esophagus bronchi trachea lungs or mediastinum Fusiform aneurysms may be found in either the thoracic aorta or in the abdominal aorta

Aneurysms may result from acute infections and these are usually small dilatations found especially in the sinuses of Valsalva They are due either (1) to implantations of infected emboli or (2) to lodgment of emboli in the vasa vasorum Occasionally they are found in the pulmonary artery due to emboli from the pulmonary valve Implantation of the infected material on the intima is usually secondary to bacterial endocarditis and may or may not be directly continuous with the vegetative endocarditis When bacterial emboli containing streptococci or staphylococci lodge in the vasa vasorum localized suppurative foci may develop in the aortic wall The lesions may be circumscribed often only a few millimeters in diameter and may bulge forming an aneurysm The intima not uncommonly ruptures leaving a stellate or linear tear This lesion is known as an acute mycotic aneurysm with rupture Bacterial stains should be done routinely in these cases

*Histologically* sections of the scarred wall of an aneurysm may fail to yield evidence of the etiology because of degenerative changes If parts of the original wall persist there may be evidences of scarring vascularization and endarteritis of the vasa vasorum indicating syphilis Sections of the aortic wall adjacent to the aneurysm commonly reveal the nature of the process whether syphilitic infectious or arte

riosclerotic The sac may be lined by thickened intima or by a laminated thrombus

108 RUPTURE OF THE AORTA (spontaneous) includes partial or complete rupture of the vessel wall in the absence of extrinsic pathological processes Spontaneous rupture never occurs in a normal vessel although on gross inspection in some instances the vessel may appear normal Medial necrosis of the aorta with or without other degenerative or inflammatory lesions and especially if accompanied by arterial hypertension is of paramount etiological significance More rarely *congenital malformations* such as *coarctation* and *hypoplasia* of the aorta may predispose to rupture The arch of the aorta 2 to 3 centimeters above the aortic valve is the most frequent site of rupture This area lies within the pericardial sac and escaping blood may readily accumulate there and lead to cardiac tamponade The tear is almost always transverse and may follow the circumference for a variable distance In other instances when the coats of the aorta are dissected by the tear and the escaping blood the picture is one of so-called dissecting aneurysm Hematoma in the adventitia of the aorta following leakage from an aneurysmal sac is not to be confused with dissecting aneurysm

109 DISSECTING ANEURYSM (*dissecting hematoma*) is most common in the ascending portion of the aorta After the blood has forced its way between the coats of the vessel dissection varying from a few centimeters to complete separation of the layers throughout the length and circumference of the aorta and even extension into its branches may occur The dissection travels through the middle third of the media as a rule Rarely a second rupture into the lumen of the aorta may occur distally creating a double barreled aorta In most instances the second rupture occurs into the adventitia with hemorrhage into the pericardial sac mediastinum pleura or retroperitoneal tissue Although death may ensue rapidly instances of healing resulting in a double barreled aorta may be encountered

*Histologically* the essential lesion is that of medial necrosis Sections incorporating the edges of the tear may not show fresh necrosis Several sections of adjacent areas may be necessary to establish the presence of medial degeneration or necrosis In some instances the medial lesion may be associated with arteriosclerosis of the vasa vasorum (especially when associated with hypertension) or in those rare cases associated with syphilitic aortitis changes in the vasa vasorum may be found In cases with dissection of the aortic wall sections of the adja

cent aorta will reveal a layer of blood in the middle third of the media either localized or extending throughout the length of the dissection

110 THROMBOSIS OF THE AORTA is encountered most frequently in the abdominal portion near the bifurcation where severe degrees of arteriosclerosis with calcification and ulceration are common Only rarely are the thrombi sufficiently large to occlude the lumen The majority are of the mural type varying from 2 or 3 millimeters to about 2 centimeters in diameter and length So called saddle thrombi are *occasionally found at the bifurcation Usually large ulcerated* atheromatous plaques are located beneath the thrombi Mural thrombi of infectious nature occur in the aorta commonly in the ascending portion They are generally small and are associated with bacterial endocarditis of the aortic valve

*Histologically* the fresh thrombus consists of platelet columns with marginated leucocytes and fibrin threads between which red blood cells are suspended In those thrombi which are only found on micro scopic examination the character is generally that of a homogeneous membrane with tinctorial properties of fibrin implanted on an atherosclerotic plaque or syphilitic aortitis Occasionally it is im possible to differentiate even histologically between an embolus in the aorta and a mural or parietal thrombus In some specimens evidences *of both may be found i e an embolus with a propagating thrombus*

111 EMBOLISM OF THE AORTA is far less frequent than thrombosis It may be considered as the probability when there is associated heart disease with intracardiac thrombi or with acute or subacute bacterial endocarditis The most common site of lodgment of the embolus is at the bifurcation

112 THROMBOSIS AND EMBOLISM OF THE PULMONARY ARTERY — *Thrombosis* may involve the main vessel or its branches and produce *partial or complete obstruction* The *thrombus is frequently* super imposed on arterial lesions either inflammatory or degenerative nota bly arteriosclerosis and also in the presence of congenital anomalies Secondary pulmonary thrombosis is common around emboli In some instances thrombi develop slowly and may undergo canalization and fibrosis Infarcts of the lung may or may not be found

*Pulmonary emboli* most frequently occur in the presence of heart diseases after surgical operations and trauma They often originate from thrombosed veins in the lower extremities or in the pelvis They

usually consist of a long thick clot either folded or in a coiled mass In all necropsies presenting an embolus or a thrombus in the pulmonary arteries especially of the main trunks a search should be made for the source particularly in the lower extremities and in the veins of the pelvis Examination of the pulmonary artery in situ is the surest way to determine the presence of an embolus or a thrombus The embolus may be found obstructing the right or left pulmonary artery riding the bifurcation filling the main trunk itself or extending into the right ventricle and atrium

There are rare instances where an embolus enters the arterial circulation through a patent foramen ovale (paradoxical embolism)

*Histologically* the pulmonary arteries may be normal or show signs of underlying disease usually arteriosclerosis and rarely syphilis

113 INJURY OF THE AORTA —a *Blunt force injuries* —The ascending portion of the aorta may be torn partially or completely by traction on the heart or a transverse rupture may be caused by sudden increase of intravascular pressure as a result of chest injury The ruptures possess uneven edges and are usually accompanied by sudden fatal hemorrhage into the pericardium or into one of the chest cavities The ascending aorta is the site of predilection for spontaneous ruptures and unless there are unmistakable indications of severe violence applied to the chest microscopic examination should be made of the aortic wall near the site of the lesion to determine the presence of a pre existing lesion

In the arch and descending portions of the thoracic aorta incomplete or complete transverse tears may occur as a result of downward traction on these vessels the most common site being just below the origin of the left subclavian artery The descending aorta is sometimes torn by fracture luxations of the thoracic vertebrae In some cases the aorta is crushed or dislocated from its bed by a grinding force The violence which produces these injuries is necessarily severe Death occurs from hemorrhage into the chest cavities

The abdominal aorta or the iliac arteries may be ruptured by severe impact on the lower abdomen causing retroperitoneal hemorrhage or hemorrhage into the peritoneal cavity

b *Penetrating injuries* Bullet wounds appear as characteristic stellate perforations of the aorta or its wall may be grazed or furrowed or even severed Stab wounds are sharp-edged and their shape depends upon the form of the blade a broad blade may sever a vessel Foreign bodies may reach the aorta through several avenues of entrance but most of them pass through the gastro-intestinal tract

Perforating wounds of the aorta or pulmonary artery cause hemorrhage into the nearest large body cavity

114 ARTERIOVENOUS FISTULA —An arteriovenous fistula may be produced by an object such as a bullet knife point or spicule of bone which penetrates a juxtaposed artery and vein forming a communicating channel between the two *This is most often seen in the extremities* but has occurred between the aortic arch and the left innominate vein and the internal carotid artery and the cavernous sinus

CHAPTER IX

# DISEASES OF THE PERICARDIUM

115 PERICARDITIS should be classified according to the duration of the process the character of the inflammatory reaction and the nature of the exudate e g acute fibrinous pericarditis Common varieties of pericarditis are *serofibrinous fibrinous fibrinopurulent suppurative or purulent organizing fibrinopurulent* and *adhesive* Inflammatory reactions limited to the epicardium without involvement of the mesothelial surfaces or parietal pericardium are sometimes designated as epicarditis

Pericarditis should also be designated by indicating the exciting cause or etiological agent e g tuberculous pericarditis Pericarditis may be due to bacterial infection or may be non bacterial in origin Non bacterial pericarditis occurs most commonly in acute rheumatic fever in uremia and following myocardial infarction

a *In serofibrinous pericarditis* the pericardial fluid is increased and slightly turbid due to flakes of fibrin The serous surfaces are dull and covered with a delicate deposit of fibrin *Histologically* eosinophilic filaments of fibrin cover the mesothelial cells The latter are often unusually well preserved although swollen

b In *fibrinous pericarditis* the fibrin deposits are abundant and the pericardial surfaces may have a granular dull yellowish-gray or a shaggy ragged appearance *Histologically* the fibrin is often compressed its fibrillar characteristics lost, and solid irregular masses of deeply eosinophilic material are noted Hyperemia is observed in the adjacent connective tissues

c In *fibrinopurulent pericarditis* the exudate contains an abundance of polymorphonuclear *neutrophils* as well as fibrin

d In *suppurative* or *purulent pericarditis* disintegration of the leucocytes in the exudate is noted

e In *organizing* types of pericarditis a zone of young capillaries and fibroblasts *together with* mononuclear leucocytes forms a layer of granulation tissue at the junction between the exudate and the underlying pericardium The newly formed blood vessels are often placed at right angles *to the* surface *The epicardial fat* is usually relatively unaltered As healing proceeds new connective tissue is laid *down*

The exudate is resorbed and fibrous adhesions of varying extent may cause permanent union between the two pericardial layers. Occasionally these may have the appearance of slender bands or synechiae. f *Chronic pericarditis,* including the adhesive type, may be associated with systemic disease as in lupus erythematosus, scleroderma, rheumatoid arthritis or polyserositis or Pick's disease. Usually the *recognition of the nature of such lesions depends upon the finding of* characteristic lesions in other organs. In lupus erythematosus, however, the fibrous tissue may *have a gelatinous appearance and may* stain metachromatically.

116 ADHERENT PERICARDIUM with obliteration of the pericardial sac by fibrous adhesions usually results from previous inflammation but *the nature of the original lesion is not always discoverable.* This lesion is sometimes referred to as *constrictive pericarditis.* It is not always possible to correlate clinical manifestations of pericardial constriction with the anatomical findings.

117 HEMOPERICARDIUM may be post-traumatic or may follow rupture of dissecting or saccular aortic aneurysms or of myocardial infarcts. *If the parietal layer is intact, free bleeding into the sac rapidly* causes fatal tamponade of the heart. Epicardial hemorrhage is common in most conditions with hemorrhagic tendencies.

118 HYDROPERICARDIUM (over 75 cc of free fluid) may occur in any condition in which tissue fluids are increased, as in congestive heart or renal failure. It is sometimes associated with mediastinal masses *such as metastatic bronchogenic carcinoma, lymphoblastoma* or aortic aneurysm. Such fluid accumulations may be due to local compression of veins or direct invasion of the pericardium.

119 ADIPOSE CHANGES IN THE PERICARDIUM.—The epicardial adipose tissue may be excessive in obese persons and cover the entire epicardium. In some instances it may even extend into the right ventricular myocardium. In wasting disease the epicardial fat may undergo serous atrophy. Such atrophic fat has a watery appearance and, histologically, the interstices between the shrunken adipose cells are filled with an eosinophilic granular protein precipitate, indicating that watery fluid has replaced the fat.

120 UREMIC PERICARDITIS usually occurs terminally in chronic renal disease in the form of an acute mild fibrinous inflammation. In more extreme lesions the exudate may be hemorrhagic and partly organized.

The inflammatory reaction is non specific histologically and the identification of the uremic origin of the lesion depends upon the demonstration of a renal lesion of sufficient extent to cause renal insufficiency or failure

121 PERICARDITIS FOLLOWING MYOCARDIAL INFARCTION is usually limited to the epicardial surface over the zone of infarction and is often fibrinous in character In extensive myocardial infarction without complete rupture a hemorrhagic exudate may cover the entire pericardium Local or diffuse fibrous adhesions form on healing

122 ACUTE NON SPECIFIC PERICARDITIS—A mild benign form is recognized clinically It occurs chiefly in young adults and is not associated with other demonstrable heart disease Although it leaves no apparent permanent injury to the pericardium recurrences are not uncommon

123 RHEUMATIC PERICARDITIS (see Chapter II) almost always occurs in conjunction with myocardial and endocardial lesions In the early stage it may be limited to the basal or atrial portions of the pericardial sac The lesion varies in intensity from mild epicarditis to diffuse *fibrinous* or *serofibrinous pericarditis* In later stages the exudate undergoes organization leading to *adhesive pericarditis* with or without calcification The histological character of the inflammatory reaction is not specific and recognition of the rheumatic nature of the lesion depends upon the finding of rheumatic lesions elsewhere

124 BACTERIAL INFECTION of the pericardial sac may be secondary to pulmonary or pleural lesions to septicemia and to mediastinitis It may follow lobar pneumonia and empyema in which case pneumococci are commonly the cause In septicemia due to many types of organisms with or without bacterial endocarditis pericarditis may develop Hemolytic streptococci are frequently the etiological agents In epicarditis due to pyogenic organisms the exudate is usually fibrinopurulent or suppurative In subacute bacterial endocarditis due to streptococcus viridans the pericardium is seldom inflamed

Mediastinitis with involvement of the pericardium may follow penetrating wounds of the chest or injury to the esophagus by foreign bodies such as chicken bones or toothpicks It may follow esophageal diverticulitis or severe esophagitis as is produced by swallowing corrosive substances It may develop secondary to descending infection originating in the neck It is sometimes associated with interstitial

emphysema Mixed bacterial infection is often found and the character of the pericarditis varies accordingly but purulent types of exudate are commonly present Grossly such exudates are often grayish green and foul smelling

A mild lymphocytic infiltration into the epicardium is sometimes noted *in conjunction with syphilis of the root of the aorta* but a specific luetic lesion of the pericardium is not recognized

125 TUBERCULOUS PERICARDITIS —The pericardium may be involved in tuberculosis by extension of pulmonary or pleural foci from caseous mediastinal lymph nodes or in the course of miliary tuberculosis *Tuberculous pericarditis* may vary from a few scattered miliary tubercles to extensive pericardial thickening caseation and obliteration of the pericardial sac by tuberculous granulation tissue Marked effusions may be encountered Healing is often accompanied by calcification and may result in constrictive pericarditis

*Histologically,* the tuberculous lesion is similar to that seen elsewhere The tubercles consist of epithelioid cells Langhan s giant cells and may undergo central caseation The tubercles can vary in size from the miliary ones to conglomerate nodules Often however the tubercles are poorly defined and may be even entirely lacking in later stages when tuberculous granulation tissue covers the entire epicardial surface The tuberculous character of the latter however can be recognized by the presence of large numbers of epithelioid cells and occasional multinuclear giant cells in the granulation tissue as well as by the presence of foci of caseation Absolute identification of the nature of the lesion however depends upon the demonstration of acid fast bacilli as other forms of granulomatous lesions may simulate tuberculosis These are however uncommon in the pericardium

126 The only FUNGUS INFECTION that involves the pericardium with any frequency is *actinomycosis* and this usually results as direct extension from a pulmonary lesion Such lesions generally consist of multiple abscesses separated from each other by granulation tissue Only localized areas of the pericardium are involved as a rule Irregular sinus tract formation leading into the adjacent mediastinal tissue and into the myocardium is frequently seen

The *histological characteristics* of the inflammatory reaction are not specific and the presence of the ray fungus discloses the nature of the lesion The latter may be distinguished from bacterial colonies by the presence of eosin staining radiations or clubs at the periphery and in

gram stains by the presence of branching gram positive filamentous structures within the mass

127 PERICARDIAL INJURIES —A violent force applied to the chest as in an automobile accident or in a fall from a building may cause fractures of the ribs and sternum which lacerate the pericardium A fracture of the sternum may tear an ovoid opening in the anterior surface of the pericardium Fractures of the ribs may produce separate circular and stellate punctures in the lateral and posterior aspects of that membrane at regularly spaced intervals to conform with the location of the fractures At the same time the heart may be injured and death may occur from hemorrhage into the chest

Compression of one side of the chest with fracture of the ribs on the same side forces the heart toward the opposite side tearing an ovoid opening in the lateral portion of the pericardium Severe bilateral shattering of the chest may be accompanied by ovoid pericardial tears on both the right and left sides of the sac Bursting ruptures of the heart and other cardiac injuries are usually associated with these lesions and death occurs as a result of hemorrhage into one or both chest cavities In rare instances the pericardial tear may occur alone and may not be accompanied by any immediate serious hemorrhage or other ill effect.

Stab wounds and bullet wounds may penetrate the pericardium and are associated in most cases with similar penetrating wounds of the heart Foreign bodies may also penetrate the pericardium and injure the heart

128 PRIMARY NEOPLASMS of the pericardium are extremely rare although tumors within the mediastinum especially bronchogenic carcinoma may invade the parietal pericardium Secondary tumors in the myocardium especially malignant melanoma may extend into the pericardium The latter are often but not always recognizable by the presence of melanin in the tumor cells

# NOMENCLATURE AND CRITERIA FOR THE DIAGNOSIS OF PERIPHERAL VASCULAR DISEASES

# NOMENCLATURE FOR DIAGNOSIS OF PERIPHERAL VASCULAR DISEASES

## DISEASES OF ARTERIES AND ARTERIOLES

### FUNCTIONAL CONDITIONS (VASOMOTOR)

#### A. Vasoconstrictor

| | |
|---|---|
| 1 Raynaud's syndrome (primary Raynaud's Disease) | 47–582 |
| 2 Raynaud's syndrome (secondary) | |
| a Traumatic vasospastic syndrome | 47–132a |
| b Neurovascular mechanisms | |
| (1) Cervical rib† | 47–031a |
| (2) Scalenus anticus syndrome† | 47–431a |
| (3) Hyperabduction syndrome† | 47–431a |
| (4) Spondylitis† | 47–685a |
| (5) Neuritis† | 47–586a |
| c Secondary to organic vascular disease | |
| (1) Arteriosclerosis† | 47–516a |
| (2) Thrombo-angiitis obliterans† | 47–51xa |
| (3) Syphilitic arteritis† | 47–51xa |
| d Secondary to Intoxications | |
| (1) Nicotine | 47–369a |
| (2) Tobacco | 47–369a |
| (3) Arsenic | 47–3114a |
| (4) Ergot | 47–367a |
| (5) Lead | 47–3112a |

Note Standard nomenclature uses various numbers in the 46 and 47 series to indicate particular arteries and 47x to indicate arterioles in general

a with vasospastic manifestation

b with vasodilator manifestation

This number is used in Standard Nomenclature

†If the cause of the vascular disease is a disease of another tissue the primary disease should be listed by its own anatomical and etiological numbers e g cervical rib 9 031

Indicate affected region of body by first three digits

| | |
|---|---|
| e Scleroderma | 17 –971a<br>111–971* |
| f Miscellaneous mechanisms | 47 – a |
| 3 Acrocyanosis | 17 –518a<br>099–518*° |
| 4 Cutis marmorata | 47 –586a<br>11 –410*° |
| 5 Vasospasm secondary to | |
| a Lesions of peripheral nerves† | 47 –586a |
| b Lesions of brain and spinal cord† | 47 –586a |
| c Thrombophlebitis† | 47 –586a |
| d Embolism† | 47 –496 1a |
| e Thrombosis† | 17 – 7a |
| f Trauma (post traumatic reflex sympathetic dystrophy Sudeck s atrophy post traumatic osteoporosis) | 17 –4 a<br>2 –100 9* |

B VASODILATOR

| | |
|---|---|
| 6 Erythermalgia primary | 17 –581b |
| 7 Erythermalgia secondary to | |
| a Polycythemia vera† | 17 –511b |
| b Arteriosclerosis† | 17 –516b |
| c Thrombo angiitis obliterans† | 17 –515b |
| d Trauma | 17 –1 b |
| e *Miscellaneous factors* | 17 – b |

ORGANIC CONDITIONS (STRUCTURAL)

A OCCLUSIVE (organic)

| | |
|---|---|
| 8 Arteriosclerosis | |
| a Atherosclerosis obliterans | 16 –952 1 |
| b Medial (Monckeberg s) arteriosclerosis | 1602–955 |
| c Combined | 16 –950 |
| 9 Thrombo angiitis obliterans | 102–930* |
| 10 Essential polyangiitis (periarteritis nodosa) | 102–931 1 |
| 11 Cranial arteritis (temporal arteritis) | 171–931 1 |
| 12 Ergotism | 17 –367 1 |
| 13 Arteritis secondary to | |
| a Infectious diseases† | 17 –1 1 |
| b Local inflammatory processes† | 17 –1 1 |

| | |
|---|---|
| 14 Hypertensive vascular disease | 460–533 |
| 15 Arteriolitis secondary to | 47x–1 4 |
| a Infectious diseases† | –1x0 4 |
| b Local inflammatory processes† | 47x–1 4 |
| c Lupus erythematosus disseminatus† | 47x–910 4 |
| d Idiopathic arteriolitis | 47x–930 4 |
| 16 Arterial Thrombosis | |
| a Associated with infectious diseases† | 46 –1 7 |
| b Associated with blood dyscrasias† | 46 –54 7 |
| c Secondary to trauma or compression† | 46 –4 7 |
| d Secondary to surgery† | 46 –415 7 |
| e Associated with parturition† | 46 –417 7 |
| f Associated with cardiac insufficiency† | 46 –519 7 |
| g Associated with slowed blood stream† | 46 –510 7 |
| h Associated with exposure to radiation | 46 –47 7 |
| i Idiopathic | 46 –619 |
| 17 Abscess of wall of artery | 46 –1 2 |
| 18 Frost bite | –448••<br>47 –448 |
| 19 Pernio | 110–446••<br>47x–446 |
| 20 Livedo reticularis | 47x–900a<br>447–5x2•• |
| 21 Arterial embolism | |
| a Thrombus† | 46 –496 4 |
| b Fat† | 46 –426 |
| c Air† | 46 –427 |
| d Bacterial† | 46 – 4 |
| e Neoplastic† | 46 –8 4 |
| f Fungus† | 46 –2 4 |
| g Inorganic substances | 46 –429 4 |

## B Non-occlusive (organic)

| | |
|---|---|
| 22 Aneurysm | |
| a Congenital | 46 –015 |
| b Syphilitic | 46 –147 6 |
| c Arteriosclerotic | 46 –912 6 |
| d Mycotic | 46 –100 6 |
| e Traumatic | 46 –4 6 |
| f Embolic† | 46 –618 6 |

| | |
|---|---|
| e Scleroderma | 17 –971a<br>114–971* |
| f Miscellaneous mechanisms | 47 – a |
| 3 Acrocyanosis | 47 –518a<br>099–518* |
| 4 Cutis marmorata | 17 –586a<br>11 –440*° |
| 5 Vasospasm secondary to | |
| a Lesions of peripheral nerves† | 47 –586a |
| b Lesions of brain and spinal cord† | 47 –586a |
| c Thrombophlebitis† | 47 –586a |
| d Embolism† | 17 –196.1a |
| e Thrombosis† | 17 – 7a |
| f Trauma (post traumatic reflex sympathetic dystrophy Sudeck's atrophy post traumatic osteoporosis) | 17 –4 a<br>2 –100.9*° |

B VASODILATOR

| | |
|---|---|
| 6 Erythermalgia primary | 17 –581b |
| 7 Erythermalgia secondary to | |
| a Polycythemia vera† | 17 –511b |
| b Arteriosclerosis† | 17 –516b |
| c Thrombo angiitis obliterans† | 17 –515b |
| d Trauma | 17 –1 b |
| e Miscellaneous factors | 17 – b |

ORGANIC CONDITIONS (STRUCTURAL)

A OCCLUSIVE (organic)

| | |
|---|---|
| 8 Arteriosclerosis | |
| a Atherosclerosis obliterans | 16 –952.1 |
| b Medial (Monckeberg's) arteriosclerosis | 1602–955 |
| c Combined | 16 –950 |
| 9 Thrombo angiitis obliterans | 102–950* |
| 10 Essential polyangiitis (periarteritis nodosa) | 102–931.1 |
| 11 Cranial arteritis (temporal arteritis) | 171–931.4 |
| 12 Ergotism | 17 –367.1 |
| 13 Arteritis secondary to | |
| a Infectious diseases† | 17 –1 .1 |
| b Local inflammatory processes† | 17 –1 .1 |

| | |
|---|---|
| (b) Myelogenous leukemia† | 18 –5137 |
| (c) Lymphatic leukemia† | 18 –5157 |
| (d) Pernicious anemia† | 18 –5127 |
| (e) Disturbances of blood clotting mechanism† | 18 –5197 |
| | 18 –5197 |
| (f) Other blood dyscrasias† | 18 –5 7 |
| (9) Cardiac insufficiency† | 18 –5197 401 |
| (10) Carcinoma† | 18 –8 7 |
| 2 Neoplastic invasion of vein | 18 –8 4 or 7 |
| 3 Venous compression by | |
| a Gravid uterus† | 18 –135 1 |
| b Neoplasm† | 48 –139 1 |
| c. Aneurysm† | 18 –135 1 |
| d Scar tissue† | 48 –135 1 |
| e Scalenus anticus syndrome† | 18 –131 1 |
| f Hyperabduction syndrome† | 18 –131 1 |
| g Fractures† | 18 –136 1 |
| h Dislocations† | 18 –131 1 |
| i Increased intra abdominal pressure† (ascites) | 48 –522 1 |

B NONOCCLUSIVE

| | |
|---|---|
| 1 Varicose veins | |
| a Primary | 18 –9x6 |
| b Secondary to | |
| (1) Posture | 18 –131 9 |
| (2) Occupation | 48 –132 9 |
| (3) Clothing | 18 –133 9 |
| (4) Proximal obstructive lesions or pressure† (See II A 3 Diseases of Veins) | 48 –522 9 |
| (5) Thrombophlebitis† | 48 –522 9 |
| (6) Arteriovenous anastomosis† | 48 –522 9 |
| (7) Hemangioma | 18 –8x0 |
| (8) Congenital anomalies of veins | 18 –010 9 |
| 2 Arteriovenous anastomosis (fistula) | |
| a Congenital | 402–029 |
| b Traumatic | 402–100 3 |
| c Secondary to malignant lesions† | 402–8 3 |
| d Secondary to bacterial infections† | 402–1 3 |
| e Secondary to fungus infections† | 402–2 3 |

| | |
|---|---|
| g Idiopathic | 16 –910 6 |
| 23 Essential polyangiitis (periarteritis nodosa) | 102–931 |
| 24 Arteriovenous anastomosis (fistula) | |
| a *Congenital* | *102–029* |
| b Traumatic | 402–4 3 |
| c Secondary to malignancy | 102–8 3 |
| d Secondary to bacterial infections | 102–1 3 |
| e Secondary to fungus infections | 102–2 3 |
| 25 Congenital anomalies of artery | 16 –0 |
| 26 *Trauma of artery* | *16 –4* |
| 27 Scalenus anticus syndrome† | 16 –134 |
| 28 Rupture of artery† | 16 –1 5<br>–1 5 |
| 29 Effects of exposure to radiation | 46 –47 |

## DISEASES OF VEINS

### FUNCTIONAL CONDITIONS (VASOMOTOR)

| | |
|---|---|
| Spasm | 48 –582<br>–4 a |

### *ORGANIC CONDITIONS (STRUCTURAL)*

#### A Occlusive

| | |
|---|---|
| 1 Thrombophlebitis and venous thrombosis (phlebothrombosis) | |
| a Primary | |
| (1) *Thromboangiitis obliterans* | *402–930** |
| (2) Migratory thrombophlebitis | 480–930 7 |
| (3) Essential or idiopathic local | 18 –900 7 |
| b Secondary to | |
| (1) Mechanical injury | 18 –4 7 |
| (2) Muscular effort or strain | 18 –13x 7 |
| (3) Chemical injury | 18 –3 7 |
| (4) Inflammatory or suppurative lesions (etiologic agent to be indicated) | 18 –1 7<br>–2 7 |
| (5) Infectious diseases† | 18 –1 7 |
| (6) Severe ischemia† | 18 –511 7 |
| (7) Varices† | 48 –522 7 |
| (8) Blood dyscrasias | |
| (a) Polycythemia† | 48 –541 7 |

# DISEASES OF THE LYMPHATIC SYSTEM

| | |
|---|---|
| | 540–854A |
| 1 Lymphangioma | 510–854A 1 |
| a Simplex | 510–854A 3 |
| b *Cysticum* | 510–854G |
| 2 Lymphangiosarcoma | |
| 3 Lymphedema | |
| a Primary | |
| (1) Congenital (Milroy's disease) | { –013* 510–013 |
| (2) Praecox | 510–900 8 |
| b Secondary to | |
| (1) Surgical removal of lymph nodes | 55 –415 8 |
| (2) Neoplastic invasion of lymph nodes | 55 –8 8 |
| (3) Lymphadenitis due to | |
| (a) X ray | 55 –4711 8 |
| (b) Pyogenic infection | 55 –1 8 |
| (c) Granulomatous infections | 55 –2 8 |
| (etiologic agent to be indicated) | –1 8 |
| (4) Dependency edema | 510 –431 8 |
| 4 Inflammatory lesions | |
| a Acute lymphangitis | 54 –1 |
| b Chronic lymphangitis | 54 –1 |

# DISEASES OF MINUTE VESSELS

## INCREASED FRAGILITY OF VESSELS

| | |
|---|---|
| 1 Infectious purpura due to | |
| a Bacterial | 190–1 5 |
| b Virus and other micro-organisms | 190–1 5 |
| 2 Toxic purpura due to | |
| a Arsenic | 190–3114 5 |
| b *Phosphorus* | 190–3123 5 |
| c Phenolphthalein | 190–31211 5 |
| d Heparin and related substances | 190–3329 5 |
| e Coumarin derivatives and related substances | 190–360 5 |
| f Venom | 190–381 5 |

| | |
|---|---|
| 6 Aberrant position of vein | 48 –021 |
| 7 Hypoplasia of vein | 48 –016 |
| 8 Phlebectasia | 48 –015 |
| 9 Periphlebitis | 48 –190 |
| 10 Phlebosclerosis | 48 –952 |
| 11 Rupture of vein | 48 –4 5<br>–1 5 |

# NEOPLASMS OF BLOOD VESSELS

## HEMANGIOMA

| | |
|---|---|
| 1 Cavernous hemangioma | 190–850A |
| 2 Capillary hemangioma | 190–850A |
| 3 Plexiform hemangioma | 190–850A |
| 4 Sclerosing hemangioma | 190–858A |
| 5 Syndromes with hemangiomas | |
| a Multiple hemangiomas and chondromas (Kast's syndrome) | 190–850A<br>–873B |
| b Hemangioma of retina and central nervous system | x23–850A<br>900–850A |

## HEMANGIO ENDOTHELIOMA

| | |
|---|---|
| 6 Hemangio endothelioma—benign | 190–850A |
| 7 Hemangio endothelioma—malignant | 190–850B |

## SARCOMA

| | |
|---|---|
| 8 Angiosarcoma | 190–850G |
| 9 Kaposi's sarcoma | 190–852 |
| 10 Ewing's sarcoma | 190–875G |
| 11 Glomus tumor (angioneuromyoma) | 190–8532 |
| 12 Hemangiopericytoma | 190–8531 |
| 13 Telangiectasis | |
| a Hereditary hemorrhagic telangiectasia | 490–851A |
| b Papillary varices | 11x–8021 |
| c Spider angioma | 110–850A* |

# CRITERIA FOR DIAGNOSIS OF PERIPHERAL VASCULAR DISEASES

## DISEASES OF ARTERIES AND ARTERIOLES

### FUNCTIONAL CONDITIONS (VASOMOTOR)

#### A VASOCONSTRICTOR

1 RAYNAUD'S SYNDROME (primary or idiopathic) PRIMARY RAYNAUD'S DISEASE —A vasospastic condition beginning usually in early adulthood and affecting women about three times as commonly as men It is usually symmetrical The small arteries and arterioles of the fingers toes ears nose or cheeks become constricted upon exposure of the body (not just the hands) to cold or under the influence of emotion The affected acral parts become pale or cyanotic and numb but usually not very painful Upon cessation of the stimulus rubor develops with tingling or a burning sensation lasting 15 or 20 minutes Oft repeated attacks may induce trophic changes and painful ulcers of the finger tips or other parts Extensive gangrene rarely develops

Localized scleroderma (sclerodactylia acrosclerosis) may eventuate the skin becoming shiny tense hard and pigmented and the joints ankylosed However few cases of Raynaud's syndrome develop this and most scleroderma is not the result of Raynaud's syndrome Calcinosis is a rare sequel Secondary Raynaud's phenomenon must be eliminated to make the idiopathic diagnosis

2 RAYNAUD'S SYNDROME (secondary) —

a *Traumatic vasospastic syndrome* (traumatic vasospastic syndrome pneumatic hammer disease)—Raynaud's syndrome may result from the use of vibrating tools This is usually unilateral at first Thereafter exposure to cold may precipitate an attack as in the idiopathic variety Recovery is not common even when the occupation is abandoned

b *Neurovascular mechanisms*—

(1) Cervical rib—Cervical ribs arising from the seventh sixth or less commonly the fifth cervical vertebra exert pressure on the subclavian artery brachial plexus or sympathetic fibers sufficiently to produce a Raynaud's syndrome frequently with paresthesias coldness and cyanosis on abduction and in rare cases enough impairment of circulation to cause gangrene

| | | |
|---|---|---|
| 3 | Purpura due to avitaminosis | |
| | a Scurvy† | 190—763 5 |
| | b Lack of vitamin K† | 190—766 5 |
| | c Other vitamin deficiency† | 190—7 5 |
| 4 | Purpura secondary to increased venous pressure† | 190—522 5 |
| 5 | Menstrual purpura† | 190—786 5 |
| 6 | Senile purpura | 190—797 5 |
| 7 | Idiopathic purpura | *507—7911 |
| | a Henoch's purpura | 190—9x7 |
| | b Schoenlein's purpura | 190—9x71 |
| 8 | Allergic purpura | 190—390 5 |

## INCREASED PERMEABILITY OF VESSELS

| | | |
|---|---|---|
| 9 | Urticaria | 190—390 8 |
| 10 | Sensitivity to physical agents | |
| | a Mechanical | 190—1 8 |
| | b Cold | 490—11x 8 |
| | c Heat | 190—119 8 |
| 11 | Hematogenic purpura due to | |
| | a Thrombocytopenia† | 190—518 5 |
| | b Leukemia† | 190—513 5<br>—515 5 |
| | c Aplastic anemia† | 190—512 5 |
| | d Granulocytopenia† | 190—511 5 |
| | e Disturbances of clotting mechanism† | 190—519 5 |
| 12 | Local inflammation† | 190—1 8 |
| 13 | Anaphylactic shock† | 190—390 8 |
| 14 | Traumatic shock† | 190—591 8 |
| 15 | Burns† | 190—111 8 |
| 16 | Frost bite† | 190—118 8 |

and intermittent claudication or rest pain suggest the primary process

(2) Thrombo-angiitis obliterans—Raynaud's phenomenon may accompany thrombo-angiitis obliterans at first affecting one or more digits

(3) *Syphilitic arteritis*—*Raynaud's syndrome* has been *reported* to accompany syphilitic arteritis of the acral parts Positive serological tests should be fortified by histologic examination

d *Raynaud's syndrome secondary to intoxications*—

(1) Nicotine
(2) Tobacco } —Smoking causes vasoconstriction of peripheral small arteries and arterioles and is detrimental in cases of occlusive vascular disease

(3) Arsenic—Arsenic is a cause of Raynaud's syndrome To prove the relationship the vasomotor symptoms and other evidences of arsenical poisoning must clear up when the arsenic is discontinued

(4) Ergot—Ergot will produce extensive vasospasm with resultant Raynaud's syndrome and even gangrene of acral parts when hemostasis results in thrombosis Coldness cyanosis and burning pain precede the dry gangrene with black mummification Other symptoms of ergotism may *occur* involving the functions of many organs Personal susceptibility *to the drug* varies greatly

(5) Lead—Lead poisoning has been reported to have *produced* Raynaud's syndrome

e *Scleroderma* (diffuse scleroderma)—This is a diffuse vascular or collagen disease Frequently the earliest sign of its presence is the development of Raynaud's phenomena in the fingers This may be present many months before the signs of the diffuse disease such as thickening of the skin telangiectasia bone absorption calcinosis and collagenous changes in the gastro-intestinal tract lungs and heart manifest themselves

f *Miscellaneous mechanisms*—Some of these may be temporary and some chronic but any of them may sometimes produce Raynaud's syndrome poor *posture* diffuse scleroderma acral scleroderma dermatomyositis essential polyangiitis calcinosis cryglobulinemia advance pulmonary tuberculosis leukemia lupus erythematosus cold allergy paroxysmal hemoglobinuria syphilis malaria polycythemia vera and arthritis In some cases it is probable that an inherent vasomoter dis

*Roentgenograms* are confirmatory A fibrous band representing a residual cervical rib may cause the syndrome but not show on x ray study

(2) Scalenus *anticus syndrome—A neurovascular syndrome produced* by contraction spasm and hypertrophy of the scalenus anticus muscle so that the subclavian artery brachial plexus or sympathetic fibers become pinched between the scalenus anticus and medius muscles Frequently there is tenderness on pressure over the scalenus muscle As a diagnostic test the injection of procaine hydrochloride around the muscle results in temporary relief of symptoms

(3) Hyperabduction syndrome—Circumduction which brings the arms together above the head with the elbows flexed or with *the plane of their long axes corresponding to that of the body* as in sleeping with the arms behind the head may cause compression and at the same time torsion of the subclavian artery and brachial plexus resulting in parasthesias and obliteration of the radial pulses In extreme cases painful gangrene may result

(4) Spondylitis—Cervical spondylitis or ruptured nucleus polyposus may produce vasospasm with resulting color and temperature changes in the hands and fingers accompanied by parasthesias and often severe pain Aggravation is produced by *extension or twisting of the neck straining* and coughing Roentgenographic evidence is helpful with the use of myelograms if indicated to diagnose the spinal deformity

(5) Neuritis—with secondary vasospasm—Any neurologic condition that results in long standing disuse may result in disturbance of the circulation resulting in changes in color and temperature Examples include Peripheral neuritis chronic anterior poliomyelitis progressive muscular atrophy syringo myelia causalgia spina bifida and hemi and monoplegia The vascular manifestations may cause persistent cyanosis pallor coldness or the Raynaud s syndrome

c. *Raynaud s syndrome secondary to organic vascular disease—*

(1) Arteriosclerosis—The Raynaud s syndrome occurs in about 10 percent of cases of arteriosclerosis obliterans In these cases pallor or cyanosis or both are produced by cold but *usually* not by emotion Numbness and aching occur during pallor burning and tingling during recovery The age of the patient plus absence of pulsations diminished oscillometric readings

and intermittent claudication or rest pain suggest the primary process

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turbance is brought to light by the aggravating condition in other cases the Raynaud s syndrome may be merely coincidental

3 ACROCYANOSIS—Acrocyanosis is a condition characterized by persistent coldness and cyanosis of the distal parts of the extremities It is often associated with profuse sweating of the volar surfaces and often moderate local edema Pressure produces a white spot which slowly disappears Spontaneous blanching ulceration and gangrene do not occur

The mechanism is probably local sensitivity to cold or a vasomotor action producing *increased arteriolar and small artery tone* Psychoneurotic people or those with neurocirculatory asthenia are often affected Most cases occur between the ages of 20 and 45 years and the sex ratio is about equal Usually the color changes persist throughout life but in later years they may lessen

4 CUTIS MARMORATA (early or mild livedo reticularis)—Cutis marmorata (marble skin) is a mild form of livedo reticularis unassociated with any other disease Dusky red mottling of the skin appears on exposure to cold and disappears in a warm environment It is most common in girls and young women The arterioles become spasmodically narrowed and the capillaries and venules dilated The feet and legs are most commonly affected occasionally the palms forearms arms and pectoral regions Coldness numbness and aching may occur

5 VASOSPASM SECONDARY TO—a *Lesions of peripheral nerves*—Vasospasm may occur as a secondary manifestation of lesions of a peripheral nerve

b *Lesions of brain and spinal cord*—This is not common or of great practical importance unless the patient is at the same time afflicted with arteriosclerosis obliterans

c *Thrombophlebitis*—Arteriospasm occasionally accompanies acute thrombophlebitis of the adjacent vein producing temporary absence of arterial pulsations with some lowering of the cutaneous temperature The leg is swollen and cyanotic rather than pale however as distinguished from acute arterial occlusion and the superficial veins are usually distended rather than collapsed

d *Embolism*—Arteriospasm is a common accompaniment of embolism to peripheral arteries and is frequently responsible for severe pain Both the artery containing the embolus and the neighboring arteries may be affected

e *Thrombosis*—Acute arterial occlusion due to thrombosis may give rise to vasospasm *in* other arteries supplying the same region Coldness pallor pain and other evidences of ischemia are present In an extreme

case this may lead to additional thrombosis in the same or in nearby vessels.

f *Trauma* (Post traumatic reflex sympathetic dystrophy)—Reflex vascular spasm may result from trauma producing the condition known as post traumatic reflex sympathetic dystrophy Osteoporosis may develop (Sudeck atrophy or painful osteoporosis) There must be a history of trauma although this may be relatively mild or of operation preceding the appearance of symptoms An early phase may be a typical Raynaud s syndrome Frequently after a suitable time has elapsed there are tenderness edema pain cold moist slightly cyanotic skin and roentgenographic evidence of osteoporosis ( traumatic segmentary arterio-spasm )

B VASODILATOR

6 ERYTHERMALGIA *primary* (erythromelalgia)—Erythermalgia is an uncommon condition in which the extremities especially the palms and soles are red hot painful and often somewhat swollen Vasodilatation is the characteristic feature The condition is aggravated by dependency and a warm environment and moderately relieved by elevation and cooling It usually occurs in middle or late life and is chronic in the primary form Arteries pulsate unusually well unless there is associated obliterating vascular disease Trophic changes ulceration and gangrene do not result from erythermalgia whether primary or secondary

7 ERYTHERMALGIA—secondary to—

a *Polycythemia vera*

b *Arteriosclerosis*—Typical erythermalgia may result from this cause More usually however it causes rubor with coldness of the skin which must not be diagnosed as erythermalgia

c *Thrombo angiitis obliterans*—The same comments may be made

d *Trauma*—In the early stage of post traumatic reflex sympathetic dystrophy erythermalgia may be present.

e *Miscellaneous factors*—It may be secondary to fever hyperthyroidism alcoholism neurocirculatory asthenia menopause hypertension diabetes mellitus gout, rheumatoid arthritis organic neurologic disease and poisoning by thallium mercury and arsenic pellagra especially in the early stages beri beri and the wearing of nylon hose In many of these secondary cases the condition clears up if the primary factor is relieved

## ORGANIC CONDITIONS (STRUCTURAL)

### A Occlusive (organic)

8 Arteriosclerosis —

a *Atherosclerosis obliterans*—This is the foremost cause of death over the age of 50 affecting as it does any or all arteries It frequently affects the extremities Pain is the chief complaint either (1) intermittent claudication typified by fatigue tightness or cramps in the calves of the legs or the muscles of the thighs after walking a limited distance relieved by rest or (2) pain at rest typically cramplike or of an aching character with the development of ulcers

*Temperature of skin* is *usually low* Sensations of coolness numbness and of pins and needles are common Atrophy of the muscles and skin with retarded growth and brittleness of the nails is an early sign

Arterial deficiency is confirmed by the development of pallor on elevation of the extremity and rubor followed by cyanosis on dependency *The venous filling time is often prolonged beyond the normal of 10 to 15 seconds* The pulsations in the accessible arteries may be diminished or absent Hardening and tortuosity may be noted on palpation Aneurysms sometimes occur in the course of an artery most often in the popliteal artery

*Ulcers* may develop on the toes heels or elsewhere often being precipitated by trauma They are usually dry but secondary infection may cause them to be moist They extend slowly are frequently undermined *at the edges and may have a black eschar at the base Gangrene* may appear as a small bluish spot gradually spreading to involve the whole toe foot or leg An entire toe may be involved at once These areas may be painless or very painful The involved portion may become mummified

Oscillometric readings help in determining the patency of major vessels and the level of their occlusion Surface temperature studies may be made but differences of less than 1 degree can be detected by the fingers *Reflex vasodilatation tests are of value in determining the presence* and degree of vasospasm Roentgenograms may show calcification and arteriograms although not essential will give information as to points of occlusion extent of collateral circulation and the presence of a true or false aneurysm

*The diagnosis* is rarely difficult but confusion with thrombo angiitis *obliterans or embolism may occur*

b *Medial* (Monckeberg s) *arteriosclerosis*—In this condition the media of the main arteries *is calcified and the lumen usually uncom*

promised It is quite common and not necessarily of serious import Pulsations are maintained Ulcers and gangrene do not occur unless atherosclerosis obliterans is also present The diagnosis is usually made by palpation of the hardened tubelike but pulsating vessels and by means of roentgenograms which show the calcification in the course of the vessels

c *Combined*—These two forms of arteriosclerosis may occur in the same individual The manifestations are mainly those produced by atherosclerosis obliterans

9 THROMBO-ANGIITIS OBLITERANS—Thrombo-angiitis obliterans (Buerger's disease) a chronic inflammatory disease of the arteries veins and secondarily of nerves occurs almost exclusively in men between the ages of 17 and 45 years It affects all races Smoking tobacco is the most important and only known etiologic factor All authentic cases are smokers and the condition almost invariably subsides with abstinence from tobacco The extremities particularly the lower ones are most commonly involved but any vessels may be affected The effects depend upon the vessels involved

Pain is usually the earliest and most prominent symptom It may appear first as intermittent claudication It is frequently *accompanied* by parasthesias of pins and needles formication and cold or burning sensations This is followed by rest pain which precedes *ulceration* or gangrene This is a burning gnawing type of pain which is worse at night and may be relieved by walking or by hanging the feet out from under the bed covers Ultimately ulcers or gangrene develop with severe and usually constant pain often worse at night and with overheating This pain persists until the lesion is thoroughly healed

*Phlebitis* superficial or deep occurs in about 50 percent of the cases it is usually below the knees and sometimes is the earliest manifestation It is frequently migratory

Temperature and color changes occur as with advanced arteriosclerosis obliterans and the pulsations in one or more arteries may be diminished or absent When infection supervenes the skin of the affected area may remain red during elevation Edema is common due to phlebitis inactivity inflammation or a combination of these factors

Trophic changes frequently ensue in the skin and nails the former becoming shiny scaly and pigmented the latter thick discolored ridged and slow-growing A slight injury or chilling may precipitate ulceration resulting in loss of a toe or even the whole foot Complicating bacterial or fungus infections frequently lead to deep fissures between the toes The ulcers and fissures are sharply defined deep

moist purulent and inflamed Gangrene may follow with loss of the part Arterial thrombosis may occur with more or less massive gangrene which is usually soft and moist After weeks and months of excruciating pain spontaneous amputation may occur

Ischemic neuritis may develop with agonizing pain described as sharp dull shooting pulling or tearing

Oscillometric readings help to determine the level of occlusion of major vessels of the extremities Arteriographic studies are of interest but rarely essential for the clinical care of the patient The presence and degree of vasospasm may be determined by tests for reflex vasodilatation sympathetic block or spinal anesthesia

In the differential diagnosis Raynaud s disease arteriosclerosis obliterans embolization thrombophlebitis acroscleroderma and the neurovascular syndrome involving the upper extremities are the main conditions to be considered

10 Essential Polyangiitis (periarteritis nodosa essential periarteritis) —

(This name is suggested because the concepts of the etiology and pathology have changed )

*Essential polyangiitis (periarteritis nodosa) is an uncommon disease* of unknown etiology but one in which hypersensitivity may play an important role It is more common in males and may occur at any age Small arteries and veins in various parts of the body are affected and produce profound effects as the result of occlusion and/or hemorrhage *The clinical course may resemble an acute subacute or chronic* infectious disease lasting for days weeks months or even years and usually ending in death with varied and bizarre local manifestations depending upon the vessels affected

Thus there may be cutaneous muscular neuritic and visceral effects In the skin and subcutaneous tissues rashes (petechial necrotic erythematous) edema and nodules (due to aneurysms or thickening of arterial walls) may occur Arthritis polymyositis and polyneuritis are common Gastrointestinal disturbances pulmonary involvement coronary arterial lesions pancreatic and hepatic dysfunction renal involvement with hypertension (in about 30 percent of the cases) and renal insufficiency and *cerebrovascular complications have all been observed*

The usual picture is that of successive exacerbations involving one area after another with malaise weakness emaciation anorexia insomnia and headaches Periods of pyrexia may alternate with long afebrile episodes Moderate or severe leukocytosis may be present and *eosinophilia* frequently occurs The sedimentation rate is usually ele

vated Anemia may be severe Often the diagnosis is made by exclusion but biopsy of an arterial lesion is the best criterion Even with biopsy the changes may not be specific for polyangiitis

11 CRANIAL ARTERITIS (temporal arteritis)—Cranial arteritis (temporal arteritis) is a rather rare febrile self limited disease of variable duration and unknown etiology involving the temporal and occasionally other cranial arteries Older persons are affected The symptoms are systemic and local The former include fever malaise sweating weakness anorexia and weight loss Locally there are pain and tenderness of one or both temporal arteries which are reddened indurated or nodular and tender The pulsations may disappear because of thrombosis Pain is often complained of in the temples or elsewhere in the head Partial or complete loss of vision sometimes occurs Delirium transient coma abnormal neurologic signs and deafness have been observed Anemia and slight leukocytosis (but not eosinophilia) may occur

Biopsy of an involved artery confirms the diagnosis Excision of the inflamed artery characteristically relieves the pain but the general symptoms continue postoperatively

12 ERGOTISM (early stages may be spastic)—Ergotism in America usually results from the improper use of drugs derived from ergot which may produce severe sympathetic stimulation with vasospasm In other parts of the world notably Europe it occurs following the use of rye bread infested with the ergot fungus

The effects are symmetrical and most pronounced in the lower extremities The nose and ears may be involved The early stage of coldness numbness and rubor or cyanosis of the limbs is reversible With continued use of the drug thrombosis of vessels and dry gangrene develop and progress to black mummification General symptoms of nausea vomiting colic diarrhea and even convulsions may occur

13 ARTERITIS SECONDARY TO—a *Infectious diseases*—Inflammatory and proliferative alterations of small arteries and arterioles occur in many infectious diseases as a secondary feature In the rickettsial diseases for instance there is widespread endothelial proliferation degeneration and necrosis of arterioles capillaries and venules with perivascular changes Thrombosis of larger arteries may occur Erythema nodosum and erythema induratum are examples of inflammatory vasculitis caused by a systemic condition

b *Local inflammatory processes*—In local inflammatory processes both acute and chronic the local arterioles capillaries and venules are involved in the tissues affected Erythema edema, ulceration and necrosis are the main manifestations and are produced by dilatation increased permeability, and thrombosis of the small vessels as the result of the infection The inflammatory process may become so extensive as to bring about gangrene of an extremity

14 HYPERTENSIVE VASCULAR DISEASE —Hypertensive vascular disease while characteristically producing severe cardiorenal effects because of arteriolar vasoconstriction terminating in organic changes in the arterioles occasionally produces discrete trophic ulcerative or gangrenous lesions of the peripheral parts of the body An area of bluish discoloration usually above a malleolus becomes a hemorrhagic bleb which breaks down and forms a superficial ulcer This slowly enlarges to a diameter of 1 to 7 cm There is little granulation or exudate and healing is slow Moderately severe pain is complained of

15 ARTERIOLITIS SECONDARY TO—

a *Infectious diseases*—As under 13 above

b *Local inflammatory processes acute or chronic*—The manifestations are similar to those seen with arteritis but less extensive

c It may be secondary to lupus erythematosus disseminatus including Libman Sacks syndrome These conditions are of unknown etiology with widespread involvement of the small arteries and arterioles Young women are most often affected

Disseminated lupus erythematosus may run an acute a subacute or chronic course with fever malaise weakness prostration and loss of weight Cutaneous lesions may occur anywhere but are most common on the face and fingers They vary from bluish to red edematous slightly indurated plaques On the face they usually cover the cheeks and bridge of the nose in butterfly configuration Exposure to sunlight aggravates the condition Purpura arthralgia and arthritis are common Pleural pericardial and peritoneal effusions may occur The lymph nodes and spleen may be enlarged Cardiac murmurs are common Azotemia and hypertension may develop but are not common Anemia leukopenia thrombocytopenia albuminuria cylindruria and microscopic hematuria are frequent laboratory findings Blood cultures are negative Bronchopneumonia is usually the terminal event after months or a few years

In more chronic cases there are remissions and exacerbations

The diagnosis of Libman Sacks syndrome should be suspected when

there are signs of progressive endocarditis with prolonged fever renal damage and negative blood cultures but the diagnosis can be made only at necropsy

d *Idiopathic Arteriolitis*—Arteriolitis not associated with recognizable disease entities is sometimes observed There have been rare cases reported of progressive disseminated arteritis and arteriolitis resembling disseminated lupus but with more prominent vascular manifestations and lacking cutaneous lesions and endocarditis Gangrene of digits and local necroses of the skin may occur

16 ARTERIAL THROMBOSIS — Arterial thrombosis may occur in a normal or diseased vessel Trauma may cause a local thrombosis or may cause thrombosis elsewhere in the body There may be a laceration or contusion in which an artery is directly injured or a contusion of soft tissues sufficient to disturb the intima and thus precipitate a thrombosis

Nontraumatic arterial thrombosis occurs in diseased vessels (see appropriate headings)

a *Associated with infectious diseases*—Infectious diseases may cause an acute arterial occlusion in two ways by direct invasion of the artery or by producing so much inflammation edema and venous stasis as to shut off the arterial supply Typhoid fever influenza and pneumonia as well as acute septic infections and ulcerative colitis may have such an effect

b *Associated with blood dyscrasias e g* — (1) *Polycythemia vera* (2) *chlorosis* (3) *anemia* (4) *leukemia*

c Secondary to trauma or compression

d Secondary to surgery

e *Associated with parturition*—Thrombosis may occur due to pressure or to reflex arterial spasm during parturition

f *Associated with cardiac insufficiency*—This association is rare and must be distinguished from embolism

g *Associated with slowed blood stream*—The following conditions may cause sufficient slowing shock a traumatic compression the hypotension secondary to hemorrhage or serious visceral damage overindulgence in narcotics long bed rest and the inactivity produced by casts and various other orthopedic appliances In patients with an arterial disease this is more likely to occur

h *Associated with exposure to x ray radium or radioactive isotopes*—Scarring and retraction may cause this syndrome Radiopaque substances when injected into arteries to produce a contrast medium sometimes produce arteritis and occlusion

1 *Idiopathic*—Arterial thrombosis sometimes occurs without apparent cause Venous thrombosis is frequently associated

17 Abscess of Wall of Artery—Abscess of the wall of an artery may arise in any of the coats It may occur from local spread of infection or may be embolic or metastatic from another focus This condition may involve many vessels of an extremity or a part of the body such as the kidney or it may be localized and nearly symptomless

18 Frost bite—Frost bite may occur from exposure to subfreezing temperatures momentarily or to a less severe temperature for a longer time It occurs more readily if the blood vessels are diseased The severe degrees of frost bite characteristically produce thrombosis of the arteries and arterioles and also involve the venous and lymphatic channels

Frost bite has been classified in four degrees depending upon its severity similar to the common classification of burns It is of military importance both in frigid climates and in the cold and anoxemia associated with high altitude flying Symptoms vary from a mild stage of erythema to gangrene Prophylaxis is extremely important When exposure to cold is complicated by wetting of the affected part or by trauma the instance of gangrene will be much greater

19 Pernio—Pernio or chilblain is due to exposure to cold and wet weather without frost bite It may be acute or chronic in type Occlusive vascular lesions occur only in the chronic stages of the disease

Immediately after exposure the skin becomes cool or cold and cyanotic with edema or blebs In the early stages there is a dermatitis which is accompanied by a cyanotic red color with slight edema The symptoms of itching and burning are made worse by exposure to heat The chronic stage is characterized by ulceration In the serious cases a hemorrhagic reaction may appear in the affected part After susceptible patients are exposed to the same cold stimulant repeatedly very painful ulcers develop leaving permanent scars with atrophy and fibrous changes This lesion has been known as leukopernio erythrocyanosis and by other descriptive terms It has been confused with erythema induratum

20 Livedo Reticularis—(Cutis marmorata)—Evidenced by blotchy discoloration of the skin of the extremities Many possible causes have been suggested including those of a faulty endocrine system It can be aggravated by cold and is associated with increased vasomotor tone The lesion may involve the skin of the hands and arms but most often

the legs and feet are affected Very rarely ulceration and even gangrene may develop

21 ARTERIAL EMBOLISM —Any foreign or abnormal particle circulating in the blood may be an embolus and cause an occlusion when it lodges in a vessel too small to permit its further passage This may occur in a diseased or normal vessel The most frequent source of arterial emboli are the mural thrombi or vegetations that form in the left side of the heart Atrial fibrillation is often present Other common causes of emboli are the shock of an operation obstetrical delivery or myocardial infarction An embolus may arise from a plaque or calcified area in a proximal artery such as the aorta A patent foramen ovale or other communication between the right and left heart permit paradoxical arterial embolism arising from clots in the venous system

The following objects may cause embolism —

a *Thrombus*

b *Fat*—Fat emboli should be suspected after trauma particularly following fractures in major bones Major operations may release fat by direct introduction into the vessel or by indirect compression Contusions to soft tissues may liberate fat particles or cause necrosis with secondary fat emboli

c *Air* Air emboli should be suspected after major trauma In addition air emboli may be produced by the opening of a large vein during injury or vascular surgery especially in the neck cardiac and thoracic surgery the introduction of air into a major vessel during a therapeutic injection transfusions or catheterization techniques

d *Bacteria*—Sepsis from focal infections blood stream infections such as pneumonia typhoid fever meningitis scarlet fever and others may produce microbial emboli These organisms may be lodged as multiple implants at various parts of the body they may agglutinate and close off small or large vessels they may lodge in heart valves or riddle the lungs or kidneys they may invade the blood vessel wall and cause secondary thrombosis or embolism Other diseases may suddenly become responsible for emboli Tuberculosis in its miliary form is an example

e *Neoplasm* The tendency of neoplasm to invade both the hematic and lymphatic systems is inherent Either or both of the systems may be involved and emboli may result

f *Fungus*—Emboli may occasionally be composed of fungus growth

h *Inorganic substances*—Such substances if injected into or near a blood vessel may become embolic

B NON-OCCLUSIVE (ORGANIC)

22 ANEURYSM—An arterial aneurysm in its primary form is simply an abnormal dilatation of an artery Trauma may injure the vessel wall The pressure of the circulating blood then causes the wall to dilate The dilatation may be diffuse or saccular The sac consists of the vessel's wall This is a "true" aneurysm

In other instances the arterial wall is *destroyed in part or completely* and the contiguous structures form a sac like enclosure through which the blood circulates This is called a "false" aneurysm which usually develops as a result of a fracture of a calcific plaque The inner wall of an aneurysm may be lined with laminated blood clots thereby causing a reduction of the pulsations

Dissecting aneurysms usually are of arteriosclerotic origin A primary tear occurs The media is destroyed by the pressure of the circulating blood and dissection progresses between the layers in either direction

a *Congenital aneurysms*—These are rare The medial coat may be undeveloped or absent dilatation may occur early in life These aneurysms are often found intracranially in the internal carotid artery or in the circle of Willis Rupture causes fatal cerebral hemorrhage

b *Syphilitic aneurysms*—Aortitis a manifestation of inadequately treated syphilis occurs most often in the ascending aorta and the aortic arch less often in the descending and the abdominal aorta and more rarely in the extremities These aneurysms may be fusiform or saccular

c *Arteriosclerotic aneurysms*—The frequency of aneurysm due to *arteriosclerosis is increasing The medial coats of arteries* affected by arteriosclerosis may rupture This may occur at an area of calcific deposit or plaque The form of dilatation of the artery depends upon the local condition of the blood vessel the contiguous structures the tension in the vessel and the degree or success of repair Mild trauma of a vessel affected by arteriosclerosis is a frequent cause of aneurysms Common examples of this are saccular aneurysms occurring in the abdominal aorta *and in the popliteal artery*

Arteriosclerosis with medial necrosis is the most common cause of dissecting aneurysms Thrombosis of the artery is a frequent complication and at times may produce complete occlusion Rupture of an aneurysm of a large artery or in a critical location may result in death Gangrene may result from aneurysm of a peripheral vessel Early diagnostic criteria are the physical findings of a pulsating mass or the

x ray appearance of enlarged vessels often with calcified walls Pain may occur if there is erosion of bone or pressure on nerves

d *Mycotic aneurysms*—The wall of the aorta or of other arteries may be weakened by a suppurative process secondary to actinomycosis tuberculosis subacute bacterial endocarditis septicemia pneumonia typhoid fever or other infectious diseases A local infection around a major vessel may so weaken it as to lead to an aneurysm or to rupture Aneurysms occur most often where there is marked stress and strain on the arterial walls e g near joints in the extremities

e *Traumatic aneurysms*—May be of the true or false variety An injury to an artery may cause its later dilatation This development may be delayed due to the pressure of the contiguous structures or because of a concomitant hematoma

f *Embolic aneurysms*—An embolus may enlarge an artery and cause a weakening of the intimal and medial layers with secondary aneurysmal dilatation

g *Idiopathic aneurysm*—In certain instances the true cause for the aneurysm cannot be ascertained

23 Essential Polyangiitis (periarteritis nodosa)—This has been described previously under 10 but is mentioned here to emphasize the fact that it may be non-occlusive

24 Arteriovenous Anastomosis (fistula)—Arteriovenous anastomosis is an abnormal communication between an artery and a vein This communication may be (a) direct such as follows a stab or gun shot wound (b) through a saccular aneurysm involving primarily one vessel (c) by an abnormal dilatation of the walls of the several vessels involved (d) by arteries and veins opening into contiguous structures

The following signs and symptoms may *indicate* the presence of an arteriovenous anastomosis

1 Enlargement of a limb or area of body surface
2 Local increase in perspiration and growth of hair
3 Increased local temperature
4 Rubor of the affected area
5 Unusual prominence of veins
6 An audible bruit over the affected area occasionally a thrill may be felt
7 Increased oxygenation of venous blood from the area This may cause the venous blood to be brighter red than from corresponding normal veins Studies for oxygen unsaturation may confirm this finding

## B Non occlusive (organic)

22 Aneurysm —An arterial aneurysm in its primary form is simply an abnormal dilatation of an artery Trauma may injure the vessel wall The pressure of the circulating blood then causes the wall to dilate The dilatation may be diffuse or saccular The sac consists of the vessel s wall This is a true aneurysm

In other instances the arterial wall is destroyed in part or completely and the contiguous structures form a sac like enclosure through which the blood circulates This is called a false aneurysm which usually develops as a result of a fracture of a calcific plaque The inner wall of an aneurysm may be lined with laminated blood clots thereby causing a reduction of the pulsations

Dissecting aneurysms usually are of arteriosclerotic origin A primary tear occurs The media is destroyed by the pressure of the circulating blood and dissection progresses between the layers in either direction

a *Congenital aneurysms*—These are rare The medial coat may be undeveloped or absent dilatation may occur early in life These aneurysms are often found intracranially in the internal carotid artery or in the circle of Willis Rupture causes fatal cerebral hemorrhage

b *Syphilitic aneurysms*—Aortitis a manifestation of inadequately treated syphilis occurs most often in the ascending aorta and the aortic arch less often in the descending and the abdominal aorta and more rarely in the extremities These aneurysms may be fusiform or saccular

c *Arteriosclerotic aneurysms*—The frequency of aneurysm due to arteriosclerosis is increasing The medial coats of arteries affected by arteriosclerosis may rupture This may occur at an area of calcific deposit or plaque The form of dilatation of the artery depends upon the local condition of the blood vessel the contiguous structures the tension in the vessel and the degree or success of repair Mild trauma of a vessel affected by arteriosclerosis is a frequent cause of aneurysms Common examples of this are saccular aneurysms occurring in the abdominal aorta and in the popliteal artery

Arteriosclerosis with medial necrosis is the most common cause of dissecting aneurysms Thrombosis of the artery is a frequent complication and at times may produce complete occlusion Rupture of an aneurysm of a large artery or in a critical location may result in death Gangrene may result from aneurysm of a peripheral vessel Early diagnostic criteria are the physical findings of a pulsating mass or the

26 TRAUMATIC—Vascular spasm from trauma has been discussed previously Trauma can also produce a non-occlusive organic response A slight injury to an artery may produce a clot which may quickly canalize and endothelialize

27 SCALENUS ANTICUS SYNDROME—This has been previously discussed but may also produce a non-occlusive organic vascular response similar to the above

28 RUPTURE—A rupture of a major artery may be due to trauma either direct or indirect Rupture may be due to an accident or to surgery It may be due to disease or to a combination with trauma With arteriosclerosis syphilis or arteritis a very mild trauma may initiate rupture

29 EFFECTS OF EXPOSURE TO RADIATION—X ray radium and radioactive isotopes have marked effects upon the vascular system These are primarily due to reaction and secondarily to scarring with retraction There may be telangiectatic changes thrombosis ulceration and vascular occlusion due to contraction of scars These changes may be progressive over a period of many years At times they are associated with the development of malignancy

8 Cardiac insufficiency may be present with enlargement of the heart

9 The pulse may be slowed by pressure applied to the affected area

10 Arteriographic studies may show the abnormal communication

11 Birth marks, hemangiomas and related vascular lesions may be associated

As the result of an arteriovenous fistula short circuiting channels are set up in which part of the blood returns to the heart before it has accomplished its function. If these channels are of sufficient total size so much blood may be returned by this short circuit that nutrition to the distal part fails. This may result in cardiac hypertrophy and with large fistulas eventually in heart failure. In small fistulas this does not occur. Arteriovenous anastomosis may be

a *Congenital*—Congenital anastomoses are much more common than has previously been believed. The possibility of arteriovenous connections should be considered when there are congenital vascular anomalies such as cavernous hemangiomata, suddenly appearing varicose veins, and some of the port wine and other vascular skin anomalies. Some *congenital arteriovenous connections* are locally malignant in that after excision new anastomosis develops with extension of the process usually proximal. Though pathologically they do not have the appearance of a malignancy they may result in the loss of the part and in some cases in the loss of life

b *Traumatic*—Gunshot or stab wounds are common causes of arteriovenous fistulas

c *Secondary to Malignancy*—Malignancy may weaken the walls of a blood vessel by direct invasion resulting in an arteriovenous fistula

d *Secondary to bacterial infection* of the surrounding tissues. Such infection may invade the walls of an artery and vein producing an arteriovenous fistula

e *Secondary to fungus infections*—Such infections may act similarly

25 Congenital Anomalies (other than fistulas)—Many unusual types of congenital abnormalities are on record. These vary from congenital absence of vessels, sometimes major vessels, for which collateral circulation is developed in unusually placed channels. These anomalies are included under non-occlusive arterial diseases because the circulation to the periphery remains intact and sufficient

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# DISEASES OF VEINS

## FUNCTIONAL CONDITIONS (VASOMOTOR)

SPASM.—Spasm is a transient contraction of the circular muscle of the wall of the vein. It may occur after direct mechanical injury of a vein and also in association with disease or injury of accompanying arteries or nerves. Passive venous spasm is associated with any condition which lowers venous blood pressure, such as elevation of the part or decrease of the vis a tergo associated with arterial occlusion or arterial spasm.

Criteria for diagnosis: Observation of the contraction of the vein, particularly on comparison with the absence of the condition in similar veins in the same part or in a companion part of the body.

## ORGANIC CONDITIONS (STRUCTURAL)

### A. OCCLUSIVE

1. THROMBOPHLEBITIS AND VENOUS THROMBOSIS (phlebothrombosis). —Thrombophlebitis is thrombosis in a vein plus a variable amount of inflammatory reaction in the wall. In some instances the inflammatory reaction is primary and the thrombosis is secondary to the endothelial injury associated with this inflammatory reaction. In other instances thrombosis is primary and the thrombus may exist as such for a variable period of time, rarely more than a few hours, before secondary inflammatory reaction begins to develop in the wall of the vein. It is during this early period that embolization is most likely to occur, although it may occur at any stage of the disease. The terms "venous thrombosis" or "phlebothrombosis" are best used to indicate a stage in the development of the disease which soon progresses to embolization or thrombophlebitis. It is not always possible, clinically or pathologically, to distinguish between lesions which are primarily thrombotic and those which are primarily phlebitic.

Criteria for diagnosis: These depend on the location of the vein involved. In superficial veins, such as the long saphenous, short saphenous, median basilic, median cephalic and external jugular, palpation of the vein reveals a solid cord, usually tender and associated with local redness of the skin. When large venous trunks, such as the iliofemoral or axillary subclavian, are involved, the diagnostic triad is (a) enlargement of the involved extremity with or without edema, (b) abnormally prominent superficial veins with or without slight to

bandages dressings casts or prolonged pressure of dependent parts on operating tables or beds may occur There may be changes in the coagulation factors of the blood It is almost certain that the thrombosis is primary The phlebitis is secondary and variable in degree

(2) Muscular effort or strain—Thrombophlebitis may be due to minor trauma to valves or compression of the vein between muscles and bones resulting from muscular effort or strain although in some cases the effort or strain may be minimal This mechanism commonly affects the axillary subclavian vein less commonly the iliofemoral vein

(3) Chemical injury—Thrombophlebitis may occur locally in a vein at or near the site of a recent injection of drugs solutions used for diagnosis or sclerosing agents

(4) Inflammatory or suppurative lesions—Thrombophlebitis may occur in the region of inflammatory or suppurative lesions and may extend from such lesions proximally into larger venous trunks The thrombus may be infected and may act as a focus for dissemination of organisms throughout the blood stream Diagnosis is based upon finding thrombophlebitis in the region of a known inflammatory or suppurative lesion and recovery of the specified infecting organism from the lesion or from the blood stream or both The etiologic factor may be any pathogenic organism including actinomyces epidermophyton and others

(5) Infectious diseases—Thrombophlebitis occurs during the course of or during early convalescence from various local or systemic infectious diseases of bacterial or virus origin The lesions of the vein do not develop at the site of the infection nor is the vein invaded by the infecting organism Thrombophlebitis of this type may complicate any of the known infectious diseases even those of mild degree and benign course It probably is due to a stagnation of the venous return because of prolonged bed rest

(6) Severe ischemia—Thrombophlebitis may develop as a complication of local severe ischemia produced by acute embolic or thrombotic arterial occlusion or as the result of extensive chronic occlusive arterial disease

(7) Varices—Thrombophlebitis develops in varicose veins with or without proximal constriction or local trauma or remote trauma such as surgical operation parturition or severe injury

moderate cyanosis of the skin and (c) tenderness in the region of the involved vein e g *Scarpa s triangle or axilla Occasionally the involved* vein can be palpated

Thrombophlebitis of the deep veins of the calf may be difficult to diagnose Lesions in this location are commonly seen as a complication of surgical operation parturition prolonged bed rest or severe injury They are characterized by a rather sudden onset of pain and deep tenderness in the muscles of the calf with or without evidence of swelling edema and congestion of the superficial veins in the involved region

Acute thrombophlebitis tends to involute partially and leave a variable amount of residual anatomic damage such as thickening of the wall obstruction of the lumen and destruction of the valves

a *Primary –Thrombophlebitis may occur without any discoverable* cause There are three main forms in which it appears

(1) With thrombo angiitis obliterans–Thrombophlebitis usually involving *superficial veins sometimes companion* veins of large arteries occurs in association with the arterial lesions of thrombo angiitis obliterans

(2) Migratory thrombophlebitis–This is similar to thromboangiitis obliterans but without evidence of arterial disease and tending to involve not only superficial veins but also medium sized and occasionally large veins in any part of the body

(3) Essential or idiopathic–Local thrombophlebitis occurring as a single or recurrent episode involving the same area

b *Secondary to–*

(1) Mechanical injury (contusion laceration surgery parturition)–

(a) At the site of injury

(b) Remote from the site of injury The greatest number of cases of thrombophlebitis which are encountered clinically are in this category The condition occurs most commonly 6 to 20 days following surgical procedures parturition or severe trauma such as fracture of bones It may develop after a longer interval of enforced bed rest or if other predisposing factors are present The causative mechanism is not clear but in almost all cases there has been trauma to tissue and some loss of blood Often relative venous stasis is associated with enforced rest in bed Compression of superficial or deep veins by

5 ARTERIOVENOUS ANASTOMOSIS (See preceding text) —

6 ABERRANT POSITION OF VEIN — A congenital anomaly usually of little importance in the periphery perhaps of great importance if affecting pulmonary veins

7 HYPOPLASIA — Obviously small and undeveloped veins as compared to other structures in the part

8 PHLEBECTASIA — Dilatation of small veins frequently associated with dilated venous capillaries This is usually noted in the skin and commonly due to over exposure to roentgen rays radium ultraviolet light or sunlight or to persistently increased venous pressure chronic venous obstruction or primary atrophic diseases of the skin Sometimes they occur without discoverable cause

9 PERIPHLEBITIS WITHOUT THROMBOSIS — A lesion occurring usually in association with local inflammatory processes and rarely diagnosable except by pathologic examination There may be severe pain in the vein without evidence of thrombosis or obstruction

10 PHLEBOSCLEROSIS — Fibrosis and calcification of the walls of veins Criteria for diagnosis are X ray demonstration of calcium or the palpation of thickening or calcification of the walls of the vein

11 RUPTURE — Complete interruption of continuity of the wall of the vein secondary to injury or sudden strain or occurring in the presence of advanced destructive disease of the wall of the vein Clinical diagnosis is usually made when ecchymosis or hematoma is found in the region of an injured vein or a vein which is known to be affected by destructive disease

(8) Blood dyscrasias—Thrombophlebitis develops in the presence of many blood dyscrasias including polycythemia leukemia pernicious anemia and disturbances in blood clotting mechanism In these cases thrombosis is primary and inflammatory reaction in the wall of the vein is often minimal

(9) *Cardiac Insufficiency—Thrombophlebitis occurs in the presence* of severe cardiac insufficiency and probably results primarily from venous stasis Thrombosis is almost certainly primary with phlebitis secondary and frequently minimal

(10) Carcinoma—Thrombophlebitis occurs in any vein or veins as a single or multiple episode in patients who have carcinoma This usually is visceral carcinoma of advanced degree The lesions in the veins are primarily thrombotic and develop in veins not invaded by carcinoma cells A thrombophlebitis especially resistant to treatment may be the first clinical sign of malignancy, particularly of carcinoma

2 NEOPLASTIC INVASION OF VEIN—Occlusion of a vein may occur as a result of direct neoplastic invasion The absolute criterion for diagnosis is the demonstration of the pathologic lesion Clinical criteria are evidence of progressive venous obstruction in the presence of known neoplastic disease in the region of the obstructed vein

3 VENOUS COMPRESSION WITH OR WITHOUT THROMBOSIS OR THROMBOPHLEBITIS—Compression of a vein The criteria for diagnosis are evidence of venous compression and distal congestion in the presence of known compressing lesions such as—

Gravid uterus neoplasm aneurysm scar tissue scalenus anticus syndrome hyperabduction syndrome fractures dislocations or increased intra abdominal pressure as with ascites

B NON OCCLUSIVE

4 VARICOSE VEINS (varices)—This consists of an enlargement and tortuosity of medium and large caliber veins Enlargement occurs in diameter and in length

a *Primary*—Varicose veins occur in the absence of any known causes

b *Secondary*—varicose veins develop in the presence of and are considered to be the result of the following factors posture occupation clothing proximal obstructive lesions or pressure congenital anomalies of veins or other causes of increased venous pressure

# NEOPLASMS OF BLOOD VESSELS

## HEMANGIOMA

Hemangioma (birthmark, et cetera)—A benign growth, often present at birth, consists of vascular spaces supported by a variable amount of fibrous stroma. Hemangiomas may be subdivided into the following types:

1. Cavernous Hemangioma.—A hemangioma which contains large vascular spaces. This fact may be ascertained by the appearance of the lesion and especially by pressure upon the lesion. It gives the sensation of a reduced resistance to pressure as compared to normal tissue.

2. Capillary Hemangioma.—A hemangioma which contains small vascular spaces. This appears as a small red spot. Pressure produces a pallor of the area.

3. Plexiform Hemangioma.—A hemangioma which is not well circumscribed due to extensive ramification of the blood vessels that often penetrate deeply into the surrounding tissue.

4. Sclerosing Hemangioma.—A hemangioma in which the blood vessels are constricted by proliferating fibrous tissue. In the end stage many of the vessels are obliterated, giving the lesion a superficial resemblance to a fibroma. The sclerosing process is only evident clinically after long continued observation.

5. Syndromes with Hemangiomas—These are rare conditions.
a. *Multiple hemangiomas and chondromas* (Kast's syndrome).
b. *Hemangiomas* of the retina and central nervous system, sometimes associated with cysts of the pancreas and kidney (Lindau's or von Hippel-Lindau's disease).

## HEMANGIO-ENDOTHELIOMA

1. Benign.

2. Malignant.—A hemangio-endothelioma, like a hemangioma, contains vascular spaces but, unlike a hemangioma, it also contains solid

quently in the vertebrae It is therefore listed under bone tumors The initial symptom usually is pain

11 Glomus Tumor (angioneuromyoma)* —This benign tumor is a hypertrophied caricature of the specialized arteriovenous anastomoses found in the periphery and which are concerned with temperature control It consists of blood vessels imbedded in smooth muscle epithelioid cells and nonmyelinated nerve fibers These benign tumors may be situated anywhere in the skin but classically they are found on the hands and feet frequently beneath the nails Typically the tumor causes paroxysms of severe radiating pain especially on pressure upon the tumor The tumor is usually small seldom measuring more than a few millimeters in diameter and when superficial is slate gray in color

12 Hemangiopericytoma —This term is used to refer to a neoplasm which resembles a glomus tumor except that the epithelioid cells are lacking Most are benign

13 Telangiectasis **—Telangiectasis may not be a true neoplasm It may be that it is a varilike dilatation of preexisting vascular channels either venules capillaries or arterioles rather than a neoplastic proliferation of new vessels

a *Hereditary hemorrhagic telangiectasis***—This is characterized by numerous dilatations of the cutaneous and mucous membrane capillaries and venules These anomalies seldom measure over 3 mm Rupture of the mucous membrane lesions occasions hemorrhages manifested by epistaxis hematuria et cetera The disease is said to be transmitted by either sex as a simple Mendelian dominant

b *Papillary varices* (senile angioma Cayenne pepper spot)—Small red compressible localized benign cutaneous swellings consisting of a single dilated vessel and occurring with increasing frequency after middle age They are strictly benign

c *Spider angioma* (spider nevus nevus araneus)—A cutaneous lesion consisting of a central pulsating arteriole from which small dilated capillaries radiate outward producing a fanciful spider like appearance The lesions often are associated with liver diseases and pregnancy but may occur independently The area may vary from a few millimeters to several centimeters

* Standard nomenclature uses Glomangioma
** Standard nomenclature uses Hemangiomatosis

nests of proliferating endothelium There are all gradations between lesions in which the proliferative endothelium is markedly anaplastic and the vascular spaces crudely formed to the opposite extreme where the vascular spaces are prominent and the proliferative endothelium less conspicuous It is *impossible to differentiate morphologically* between hemangio endotheliomas which metastasize and those which are locally invasive

*Clinical manifestations* depend upon the organ involved When it involves the skin there is a circumscribed tumor usually bluish and usually compressible It may be tender or painful or both The size varies from a few millimeters to several centimeters

## SARCOMA

8 ANGIOSARCOMA *—This term is seldom used Many pathologists reserve the term sarcoma for neoplasms arising from mesenchymal derivatives other than blood vessels If many of the neoplastic cells are anaplastic but in places sufficiently differentiated to form vascular spaces the pathologist makes the diagnosis of malignant hemangio endothelioma rather than angiosarcoma Some sarcomas are so highly undifferentiated that the pathologist cannot be certain of the parent tissue

9 KAPOSI S SARCOMA **—This term is applied to multiple pigmented *lesions composed of dilated capillaries fibroblastic proliferations* inflammatory cells and hemosiderin deposits The hemosiderin deposition is due to extravasations of blood from ruptured capillaries

The lesions usually appear first *in the skin of the extremities* There is a bluish mass of blood vessels extending into the adjacent tissues and of slow growth Males are more commonly affected than females Autopsies have demonstrated that visceral lesions do occur but they are usually clinically undetectable The average life expectancy is said to be 5 to 10 years It is not at all certain whether this entity is really a sarcoma Some pathologists believe that it is essentially an inflammatory lesion

10 EWING S SARCOMA —It is widely held that Ewing s sarcoma is essentially an endothelioma and indeed the tendency of the neoplastic cells to surround spaces does suggest an endothelial origin However Ewing s sarcoma always arises in the skeletal system fre-

*Standard nomenclature uses Hemangiosarcoma
**Standard nomenclature uses Multiple Hemorrhagic hemangioma of Kaposi

# DISEASES OF MINUTE VESSELS

Not only the capillaries but also the finest arterioles and venules are included under this term

As the blood passes through the capillary network there is a constant interchange of certain of its constituents with the tissue fluids In conditions leading to increased pressure within the lumen or to damage to the capillary walls additional serum and formed elements may escape When there is an extravasation of cells into the skin or mucous membranes the term petechiae is used for minute hemorrhages which are often multiple of 2 mm size or less and purpura for larger hemorrhages Normally cutaneous hemorrhages of very slight degree occur with the constriction of a limb These are probably due to increased capillary pressure plus the added factor of anoxemia of the cells of the capillary walls

## INCREASED FRAGILITY OF VESSELS

1 Infectious Purpura—a *Bacterial*—Petechiae are characteristic of bacteremias and septicemias notably in subacute bacterial endocarditis Any overwhelming bacterial infection may give rise to capillary damage which is reflected clinically by the appearance of hemorrhages in the skin and mucous membranes of pinpoint size or large enough to be called purpura

b *Virus or rickettsia*—Purpura is characteristic of small pox and many viral infections It is found nearly always in typhus and other forms of rickettsial diseases

2. Toxic Purpura may be caused by various poisons such as

a *Arsenic*—Purpura may follow the administration of arsphenamine derivatives Fowler s solution and the like

b *Phosphorus*

c *Phenolphthalein*—A drug found in many proprietary and patent laxatives

d. *Heparin* (extract of mast cells) and related substances e g paritol and treburon

e *Coumarin derivatives and vitamin K blocking agents* (e g bis hydroxycumarin (dicumarol) ethyl biscumarin acetate (tromexan) cyclocumarol phenylindanedione and others—Slight trauma produces

genital non painful lymphedema involving all or part of one or two extremities It may be unilateral involving the upper *and lower extremities and rarely additional areas may be* involved

(2) Praecox—This refers to a clinical syndrome affecting young females much more often than males The lymphedema *usually appears between the ages of 15 and 35 involving* first the feet later ascending to involve the legs and thighs

b *Secondary Lymphedema*—is due to

(1) Surgical removal of lymph nodes

(2) Neoplastic invasion of lymph nodes either by primary or metastatic neoplasm

(3) Lymphadenitis following

(a) X ray treatment—Fibrosis and scarring enhanced by x ray may cause obstruction of the lymphatics and resultant edema

(b) Pyogenic Infection

(c) Granulomata due to

(a) Filariasis

(b) Lymphogranuloma venereum

(c) Tuberculosis

(d) Syphilis

(4) Dependent edema—*Edema of the legs may follow prolonged* sitting as during long trips by auto rail or air It has been reported in persons seated for many hours in air raid shelters Some complicating factor is usually present such as a girdle *that folds and binds the groin when seated or a chair with* a firm edge that presses on the thighs

If the edema is pale the obstruction is lymphatic If it is cyanotic a complicating phlebitis may be present

4 INFLAMMATORY LESIONS OF LYMPH CHANNELS —

a *Acute lymphangitis*—characterized by red streaks extending proximally on the extremities usually accompanied by fever and leucocytosis *They usually start from* a site of local infection

b *Chronic lymphangitis*—is associated with dull red or brownish streaks with a tendency to recurrence Etiology should be indicated if known

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purpura in patients receiving significant amounts of anticoagulant drugs In overdosage large hematomas may form

f *Snake venom insect venom*—Certain types of venom are toxic to capillaries (thrombogenic and hemolytic)

3 Purpura Due to Avitaminosis—a *Vitamin C*—Scurvy due to lack of Vitamin C is characterized by easy bruising and bleeding gums from capillary oozing Apparently the defect is in the capillary itself Sub clinical scurvy may be detectable only by the capillary fragility test and confirmed by finding a subnormal concentration of Vitamin C in the blood Specifically it is the thermolabile Vitamin C which is lacking

b *Vitamin K*—Lack of Vitamin K *produces a deficiency of pro* thrombin This results in hemorrhage from the capillaries

c *Other vitamins*—It is claimed that Vitamin P deficiency may cause capillary fragility but this is controversial

4 Purpura Secondary to Increased Venous Pressure—Increased venous pressure leads to an increase in capillary pressure Small hemor rhages may result from rupture of these vessels It is frequently seen *with defective venous return because of venous valvular defects* me chanical pressure on veins as from pelvic tumors or ascites or pro-longed dependency of the legs as in long trips by air rail or auto mobile (*especially while wearing constrictive garments such as girdles*) Garments may be responsible for increased venous pressure with sec ondary hemorrhages Such purpura are usually small in size and located in the feet and hips and about the ankles

5 Menstrual Purpura—This is a rare phenomenon of unknown cause It may occur in any part of the skin of the body except the face

6 Senile Purpura—Purpura occurs in the elderly without adequate etiological explanation

7 Idiopathic Purpura—a *Henoch s purpura*—A recurrent malady characterized by purpuric areas which may ulcerate The skin and mucous membranes are involved Attacks of acute abdominal pain may result from intestinal lesions

b *Schoenlein s purpura*—Purpura with joint manifestations The diagnosis depends upon purpura appearing in the skin and associated

with pains in the joints This may be due to hemorrhages in the joint capsules

8 ALLERGIC PURPURA — This occurs with or without thrombocytopenia.

## INCREASED PERMEABILITY OF VESSELS

Normally the capillary walls retain the solids of the blood and certain dissolved and suspended material of high molecular weight The capillary wall acts as an osmotic membrane allowing fluids and electrolytes to flow freely in accordance with physical laws Under certain conditions the capillary wall may be changed so as to allow protein and other constituents to flow through

9 URTICARIA AND ANGIONEUROTIC EDEMA — This is a result of a localized capillary dilation and edema of tissue spaces due to local action of histamine substances

10 SENSITIVITY TO PHYSICAL AGENTS — This results in local swelling due to extravasation of fluid into the tissue spaces of the skin It may appear pale or red and may be due to

a Mechanical irritation (scratching, pressure friction)
b Cold (allergic release of histamine)
c Heat (thermal damage to capillary walls)

11 HEMATOGENIC PURPURA — This is not a primary disease of the minute blood vessels They are affected by certain blood conditions so as to cause purpura The defect in at least some of these conditions is a decrease of thrombocytes It is assumed that small holes are constantly being produced in the capillaries due to trauma or loss of intercellular cement substance These holes are normally plugged by blood platelets If there is a defect in their number or chemotaxis the holes are not sufficiently plugged and hemorrhage ensues The causative conditions are as follows

Thrombocytopenia myeloid or lymphatic leukemia aplastic anemia granulocytopenia and disturbances of clotting mechanism such as hypoprothrombinemia

12 LOCAL INFLAMMATION — A change in capillary permeability is characteristic of inflammation of all types

purpura in patients receiving significant amounts of anticoagulant drugs In overdosage large hematomas may form

f *Snake venom insect venom*—Certain types of venom are toxic to capillaries (thrombogenic and hemolytic)

3 PURPURA DUE TO AVITAMINOSIS—a *Vitamin C*—Scurvy due to lack of Vitamin C is characterized by easy bruising and bleeding gums from capillary oozing Apparently the defect is in the capillary itself Subclinical scurvy may be detectable only by the capillary fragility test and confirmed by finding a subnormal concentration of Vitamin C in the blood Specifically it is the thermolabile Vitamin C which is lacking

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6 SENILE PURPURA—Purpura occurs in the elderly without adequate etiological explanation

7 IDIOPATHIC PURPURA—a *Henoch s purpura*—A recurrent malady characterized by purpuric areas which may ulcerate The skin and mucous membranes are involved Attacks of acute abdominal pain may result from intestinal lesions

b *Schoenlein s purpura*—Purpura with joint manifestations The diagnosis depends upon purpura appearing in the skin and associated

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13 ANAPHYLACTIC SHOCK may be associated with wheals or grave urticaria or edema

14 TRAUMATIC SHOCK —The picture of shock may be accompanied by the appearance of urticarial wheals sometimes blisters

15 BURNS —May cause massive exudation and loss of fluid into the tissue spaces

16 FROST BITE —Discussed elsewhere

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